CONGENITAL ABSENCE (AGENESIS) OF THE INTERNAL CAROTID ARTERY

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ABSTRACT
Agenesis of the internal carotid artery (ICA) is a rare congenital anomaly. Most of the patients are asymptomatic and it is usually discovered incidentally by computed tomography (CT) or magnetic resonance imaging (MRI). While collateral blood flow may allow these patients to remain asymptomatic, there is close association of the intra-cranial aneurysms and subarachnoid hemorrhage with ICA agenesis. Hence, recognition of this anomaly has important implications during planned carotid or transsphenoidal surgery, in thromboembolic disease, and in the surveillance and detection of associated cerebral aneurysms. We present 3 cases of absent ICA, suspected due to absent bony carotid canal on non-contrast CT (bone window settings). Out of these, only 1 patient had previous episode of hemiplegia (CVA), while the other two had unrelated symptoms. These patients were further evaluated by CT cerebral angiography, which revealed ICA agenesis with various patterns of collateral circulation. One of the patients had a co-existing developmental venous anomaly (DVA). None of our patient had sub-arachnoid hemorrhage.

KEYWORDS: Agenesis, internal carotid artery, CT angiography.

INTRODUCTION
Agenesis of the internal carotid artery (ICA) is a rare congenital anomaly with an incidence of 0.01%.¹,² The term ‘absence’ has been chosen to incorporate agenesis, aplasia, and hypoplasia of the ICA. ICA agenesis was first presented by Tode in 1787, on a postmortem examination.³ In 1954, Verbiest reported a case with ICA agenesis recognized by cerebral angiography.⁴ The etiological basis for of the carotid agenesis is not known, but may be secondary to an insult to the developing embryo.⁵

Most patients are asymptomatic, since collateral blood flow provides sufficient cerebral circulation. The most common type of collateral flow is through the circle of Willis, followed by collateral flow provided via persistent embryonic vessels or from trans-cranial collaterals originating from the external carotid artery (ECA) system.⁶

Patients may later present with subarachnoid hemorrhage, transient ischemic attack, or cerebrovascular insufficiency. There is a close association with ICA agenesis and intracranial aneurysms.⁷,⁸

Slightly more than 100 cases of congenital absence of the ICA have been reported in the literature.⁹

MATERIAL AND METHODS
Patients were selected with absent bony carotid canal on non-contrast CT (bone window settings). After taking consents and normal creatinine, patients were subjected to CT cerebral angiography on 64 slice Multidetector CT. MPR, MIP and volume rendered techniques were used for the evaluation of CT angiography.

We report case series of 3 patients with ICA agenesis with different patterns of collateral circulation.

CASES

Case 1
A 23-year-old man presented with history of tinnitus, decreased hearing and running nose. There was no significant past history. Patient underwent CT temporal bone. It revealed absence of the left bony carotid canal of petrous bone. Right petrous bony ICA canal was larger with proximally dehiscent carotid wall into the hypotympanum. Also there was pansinusitis. Both external, middle and internal ears were unremarkable.
Further CT Neck and Cerebral angiography was performed for the evaluation of left ICA. It revealed absent cervical, petrous and cavernous segments of left internal carotid artery. The left CCA was seen continuing as left ECA. Left CCA was also narrow in diameter as compared to the right CCA.

Collateral circulation was seen established by the intracranial collaterals. Intracranial left ICA was seen arising from the right supraclinoid ICA and crossing the sella and suprasellar cistern anterior to the pituitary stalk. In the medial most part of the left Sylvain fissure, it gave the left ophthalmic artery and then bifurcated into left MCA and postero-medially prominent left PCOM.

Also seen was fusiform dilatation of the right supraclinoid ICA, proximal M1 segment of right MCA and proximal A1 segment of the right ACA. There was severe near-complete luminal narrowing of the midpoint of the basilar artery over length of 9.0 mm.

Brain parenchyma was otherwise unremarkable.

Case 2
21 years old non-hypertensive, non-diabetic male patient presented with complains of chronic headache since 2 months. He also had h/o weakness of left side of body and slurring of speech, 6 months back.

CT brain plain was done, which revealed chronic infarct in the right gangliocapsular region. Bone window setting revealed absent right bony carotid canal of petrous ICA.

Further CT Neck and Cerebral angiography was performed for the evaluation of right ICA. It revealed absent right internal carotid artery in its entire extent (cervical, petrous, cavernous and supraclinoid segments).

Right CCA was seen continuing as right ECA. Right CCA was narrow in diameter as compared to the left CCA. In addition, the left ICA was seen to have an aberrant course.

Collateral circulation was established by the fetal-type collaterals. Collateral flow to the right anterior cerebral (ACA) artery via a patent anterior communicating artery (ACOM) and collateral flow to the right middle cerebral (MCA) artery via a fetal posterior communicating artery (PCOM).

The patient with pharmacologic management has remained asymptomatic for headache.

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**Fig 1 – Case 1:**

* A: Axial CT temporal bone showing absence of left bony carotid canal. *B and C:* CT cerebral angiography (axial) reveals intra-cranial left ICA arising from the right supraclinoid ICA and traversing across the sella. *D:* CT cerebral angiography (coronal) reveals narrow left and prominent right CCA, with non-visualization of left ICA. *E and F:* Volume-rendered images showing intra-cranial left ICA arising from the right supraclinoid ICA and traversing across the sella.
Fig. 2 - **Case 2:** 

A: CT Brain (Plain) reveals chronic infarct in right gangliocapsular region. 

B: CT Brain bone window settings reveal absent right bony carotid canal. 

C: CT cerebral angiography (axial) and 

D: (coronal) reveal absent entire right ICA with aberrant course of left ICA

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Fig. 3 - **Case 2:** 

E and F: CT cerebral angiography (axial) reveals absent right ICA, with collateral flow to right ACA via patent ACOM and collateral flow to right MCA via prominent right fetal PCOM. 

G: CT Brain bone window settings reveal absent right bony carotid canal. 

H: Volume-rendered CT reveals absent right ICA with collateral flow to right ACA via patent ACOM and collateral flow to right MCA via prominent right fetal PCOM.
Case 3
A 40 yrs old man with h/o schizophrenia and alcohol abuse on treatment presented with complaints of imbalance, and single episode of seizure. Patient underwent CT brain for further evaluation. Brain parenchyma was unremarkable. However, there was absent bony carotid canal on the right side.

Patient underwent CT neck and cerebral angiography. It revealed not visualization of right ICA along its entire extent, with right CCA continuing as right ECA. Left CCA and ICA were prominent along their entire extent. In addition, four-vesseled aortic arch was noted, with aberrant right subclavian artery.

Collateral circulation was established by the fetal type collaterals. Collateral flow to the right anterior cerebral (ACA) and middle cerebral (MCA) arteries via a patent anterior communicating artery (ACOM).

Also seen was a developmental venous anomaly (DVA) in the right frontal region, traversing superiorly along the right high frontal cortex and draining into the mid portion of the superior sagittal sinus.

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Fig. 4 - Case 3: A: CT Brain bone window settings reveal absent right bony carotid canal. B and C: CT cerebral angiography (axial) reveal absent right ICA, with patent ACOM supplying the right ACA and right MCA. D: CT cerebra angiography (coronal) reveals narrow right CCA continuing as ECA, with non-visualization of right ICA. E: Volume-rendered CT reveals absent right ICA with with patent ACOM supplying the right ACA and right MCA. F: CECT Brain sagittal (with positive recon) reveals co-existing DVA in right frontal region.
Few cases of absent ICA have been reported in children, suggesting that initially the collateral pathways are sufficient to support cerebral perfusion.\textsuperscript{[11]} While many cases of absence of the ICA may remain asymptomatic and go unrecognized, these patients may present later in life with symptoms related to cerebrovascular insufficiency. The entire anterior circulation may be dependent upon a single carotid artery or the vertebrobasilar system, either of which may be compromised by atherosclerosis. Congenital absence of the ICA was discovered in one of three patients during evaluation of symptoms ultimately attributed to cerebrovascular episode. Alternatively, such patients may present with mass effect from the enlarged collaterals, complications related to aneurysm, and, rarely, congenital Horner’s syndrome.\textsuperscript{[18]} The estimated prevalence of cerebral aneurysms in association with absence of the ICA is 24% to 34%.\textsuperscript{[12]} Increased flow through collateral vessels and altered flow dynamics are cited as plausible explanations for this increased prevalence.\textsuperscript{[12]} ACOM (a common collateral vessel) is the most frequent site of aneurysm formation in such cases.\textsuperscript{[19]} The increased risk of aneurysm has been listed as an indication for clinical and radiologic surveillance in these patients.\textsuperscript{[12]} There is also increased incidence of the other vascular anomalies like four vessel arch with aberrant right subclavian artery as in our case. Also, aberrant course of ICA was seen.

Recognition of this anomaly becomes important in thromboembolic disease, as emboli in one cerebral hemisphere may be explained by atherosclerotic disease in the contralateral CCA or vertebro-basilar system. Consideration of this anomaly be of utmost importance when planning carotid endarterectomy, as both cerebral
hemispheres may be dependent upon the atheromatous carotid. Identification of this variant may help prevent the erroneous diagnosis of carotid dissection or high-grade carotid stenosis, as a case of ICA hypoplasia has been mistaken for the angiographic string sign associated with a critical carotid stenosis.[17] Finally, failure to recognize the intercavernous collateral can have grave implications during trans-sphenoidal hypophyseal surgery.[20]

DISCUSSION
As Tode is credited with the first documented case of carotid agenesis, discovered on postmortem examination in 1787[16], Verbiest in 1954 reported the first case of ICA agenesis at cerebral angiography.[9] Lie[11] defined agenesis as complete failure of an organ to develop, aplasia as lack of development, and hypoplasia as incomplete development of the organ.[12] But an exact cause of these developmental anomalies has not been established, all three variations are thought to represent the sequel from an insult to the developing embryo. Postulated causes of unilateral absence have centered on mechanical and hemodynamic stresses placed on the embryo. To date, an explanation for bilateral absence has not been rendered.[13]

Lie[11] described six pathways of collateral circulation in association with absence of the ICA (Fig 5). Type A: (Fig 2 and 3 – Case 2) ---- unilateral absence of the ICA is associated with collateral circulation to the ipsilateral ACA through a patent ACOM and to the ipsilateral MCA from the posterior circulation through a hypertrophied PCOM.

Type B: (Fig 4 – Case 3) -- pattern of collateral flow, the ipsilateral ACA and MCA are supplied across a patent ACOM.

Type C: ----- represents bilateral agenesis of the ICA with supply to the anterior circulation via carotid-vertebrobasilar anastomoses, and review of the literature indicates this is generally accomplished through hypertrophy of the PCOM.

Type D: (Fig 1 – Case 1) ----- represents unilateral agenesis of the cervical portions of the ICA with an intercavernous communication to the ipsilateral carotid siphon from the contralateral cavernous ICA.

Type E: ---- diminutive ACAs are supplied by bilateral hypoplastic ICAs, and the MCAs are supplied by enlarged PCOMs.

Type F: ---- pattern provides collateral flow to the distal ICA via trans-cranial anastomoses from the internal maxillary branches of the ECA system, the so-called rete mirabile.

Others[14, 15] have simplified Lie’s original six collateral pathways into three main types: collateral flow through the circle of Willis (most frequent), collateral flow via persistent fetal circulation, and reconstitution of the ICA through skull base collaterals from the ECA. While the literature supports a nearly 3:1 left-sided predominance of ICA absence[13], in out of 3 cases 2 were right sided and one was left sided. Congenital absence may be unilateral or bilateral, although the unilateral variety is distinctly more common.[16] There may be complete absence of the ICA, or only a portion of the ICA may be missing. In our cases there is unilateral, complete absence of the ICA was seen.

Evaluation of the skull base for the presence or absence of the carotid canal is important for distinguishing aplasia from agenesis, as presence of the ICA (or its precursor) is a prerequisite for development of the carotid canal at 5 to 6 weeks of gestation. Therefore, demonstrating an absence of the carotid canal with skull base CT will confirm the diagnosis of agenesis.[12] Similarly, demonstrating the presence of a diminutive carotid canal with CT will permit one to differentiate hypoplasia of the ICA from acquired conditions resulting in a small-caliber ICA.

Acquired causes of an ICA of diminished caliber include chronic dissection, fibro muscular dysplasia, and severe atherosclerosis.[17]

CONCLUSION
ICA agenesis is a rare vascular anomaly and is most often discovered incidentally as patients are often asymptomatic. The main collateral pathways recruited in this setting include the circle of Willis, persistence of embryonic vessels, and transcranial collaterals via the ECA. Although many of these cases remain asymptomatic and go undetected, their recognition is of more than trivial interest. Congenital absence of the ICA is associated with a higher incidence of aneurysm formation. Patients may present with cranial nerve deficits, cerebral ischemia, or subarachnoid hemorrhage secondary to complications of associated aneurysm formation.

In addition to associated cerebral aneurysms, these anomalies have important implications during various neuro-intervention procedures and in the setting of thromboembolic disease. The diagnosis of ICA agenesis (distinguishing it from ICA stenosis or occlusion) and the collateral circulation should be well documented by means of CT or DSA angiography, particularly prior to carotid endarterectomy, transsphenoidal hypophyseal surgery or inducing hypotension.

REFERENCES


