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Case Report

Congenital Absence of Unilateral Internal Carotid Artery with an Intracerebral Aneurysm

Abstract

The absence of the internal carotid artery (ICA) is a rare congenital anomaly. Diagnosis of this entity is important because of its association with the cerebral aneurysm and also indicated during planned carotid or transsphenoidal surgery in thromboembolic disease and in the surveillance and detection of associated cerebral aneurysms. We report a case of congenital absence of unilateral ICA with associated cerebral aneurysm of the anterior cerebral artery.

Keywords: *Cerebral aneurysm, congenital absence of the internal carotid artery, hypertrophied posterior communicating artery*

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Introduction

Agenesis, aplasia, and hypoplasia of the internal carotid artery (ICA) are rare congenital anomalies, occurring in <0.01% of the population.[1,2] The term absence indicates agenesis, aplasia, and hypoplasia of the ICA. In the setting of an absence of ICA, the most common type of collateral flow is through the circle of Willis. Other less common collaterals are collateral flow through persistent embryonic vessels or from transcranial collaterals originating from the external carotid artery (ECA) system. Slightly more than 100 cases of congenital absence of the ICA have been reported in the literature.[3]

Case Report

A 60-year-old male presented with a headache for few months. Carotid Doppler revealed high-grade atherosclerotic stenosis of the proximal right ICA with poststenotic parvus–tardus waveforms [Figure 1]. Left carotid vessel showed high-resistance waveform with the absence of normal ICA waveform which suggested agenesis of the left ICA [Figure 1].

The patient underwent computed tomography (CT) angiography imaging of the carotid vessels and circle of Willis, which showed absent ICA on the left side and collateral flow to the left hemisphere through the circle of Willis [Figures 2-5]. The absence of the left carotid canal was also noted at bone window setting of CT, which confirmed the congenital nature of the nonvisualization of left ICA [Figure 3]. Maximum intensity projection reconstruction revealed that the left middle cerebral artery (MCA) was supplied by the basilar artery through a dilated left posterior communicating artery (PCOM) [Figures 4 and 5].

The patient subsequently underwent magnetic resonance (MR) imaging of the circle of Willis, which showed the absence of flow-related signal intensity within the left ICA [Figure 3]. Saccular aneurysmal dilatation of the anterior communicating artery (ACOM) was detected on CT angiography (CTA) and MR angiography [Figures 4 and 5]. A1 segment of the left anterior cerebral artery (ACA) and right PCOM were not seen opacified on CTA and showed the absence of flow-related signal intensity suggested aplasia [Figure 5]. The remaining portion of left ACA was supplied by ACOM aneurysm.

Discussion

Agenesis of the ICA is a rare congenital vascular anomaly having an incidence of 0.01%.[1,2] In 1787, Tode was first to report the case of ICA agenesis on postmortem examination, and a number of cases of ICA agenesis have been reported until now.[3] In 1954, Verbiest recognized

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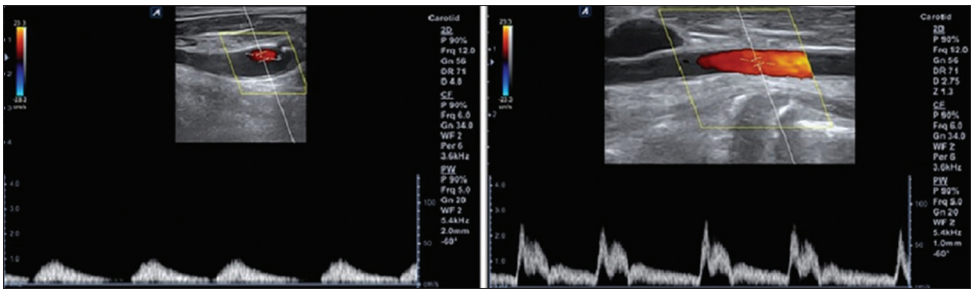

Figure 1: A 60-year-old Male with high-grade atherosclerotic stenosis of proximal right internal carotid artery and parvus–tardus waveforms in mid internal carotid artery (right image), left carotid vessel shows high-resistance waveform with the absence of normal internal carotid artery waveform suggests agenesis of left internal carotid artery

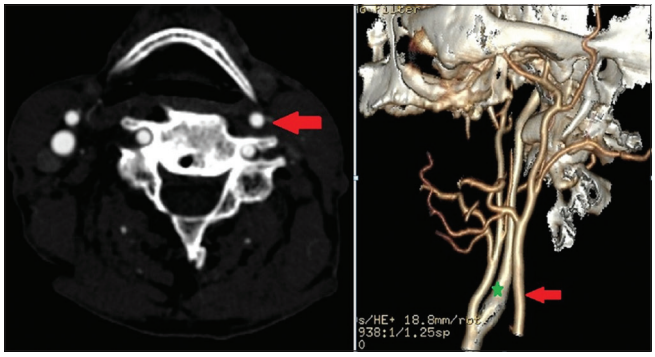

Figure 2: Computed tomographic angiography axial and three-dimensional images of the patient show nonvisualization of the left internal carotid artery (red arrows), note the normal internal and external carotid arteries are seen on contralateral side

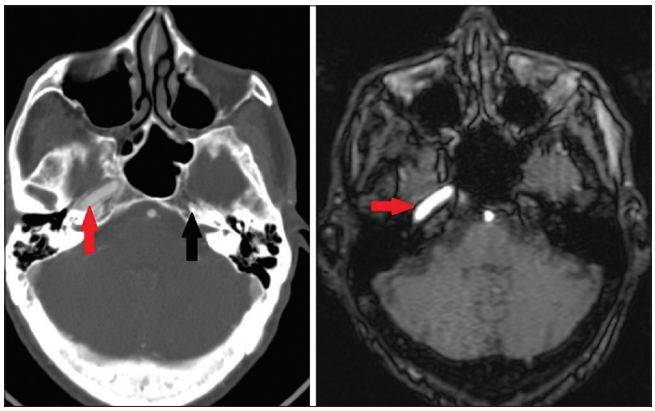

Figure 3: Computed tomographic angiography axial image of the patient shows nonvisualization of the left internal carotid artery in the carotid canal region with absence left carotid canal (black arrow) and magnetic resonance angiography shows absence of flow-related signal intensity within the left internal carotid artery, note the normal visualization and flow-related signal intensity on contralateral side (red arrows)

a case with ICA agenesis at cerebral angiography.[4] Most patients of ICA agenesis are asymptomatic because the circle of Willis provides sufficient collateral blood supply to the affected side of brain parenchyma. Later, patients may present with subarachnoid hemorrhage from aneurysmal rupture or transient ischemic attack due to vascular insufficiency.

Lie[5] defined agenesis as a complete failure of an organ to develop aplasia as lack of development with its existing precursor and hypoplasia as an incomplete development of an organ. Padget[6] described that ICA arises from the dorsal aorta and the third aortic arch at 4–5 mm embryonic stage and entirely develops by 6 weeks. Thus, agenesis of the ICA depends on involution of the third aortic arch and the distal portion of the dorsal aorta at this stage.[7,5] The origin of the common carotid artery (CCA) and ECA remains controversial. Some authors described that both the proximal ICA and the ECA arise from the third aortic arch and postulated that agenesis of the ICA should be associated with the absence of the ipsilateral ECA.[8,5] Others described that the ECA and CCA arise from the aortic sac independently and can present normally in a case of ICA agenesis.[8,5] The latter theory is more acceptable as numerous cases of ICA agenesis have been reported in the literature with normally developed ECA as was in our case.

It has been described that the carotid canal develops in association with the ICA, and the skull base does not begin to form until the 5th–6th weeks of fetal life.[9,10] Thus, if the embryonic primordium of the ICA does not develop or fails to develop before the 3rd and 5th embryonic weeks, the ICA and the carotid canal cannot develop.[9,10]

Collateral circulation accompanying ICA agenesis is classified into three forms: (1) through the circle of Willis, (2) persistent embryonic vessels, and (3) transcranial anastomosis from the ECA.[11] Tsuruta and Myazaki[12] proposed three types of collateral channels through the circle of Willis. In Type I, the ipsilateral ACA is supplied by the contralateral ICA, opposite to the ICA agenesis, through the anterior communicating artery (ACoA). The MCA is supplied by the basilar artery through the posterior communicating artery. In Type II, the ipsilateral ACA and MCA are supplied by the contralateral ICA through patent ACoA. In Type III, the ipsilateral ACA and MCA are supplied by the transcranial anastomoses that develop from ECA or contralateral ICA or primitive vessels. In our case, Type I anastomosis was determined by CT and angiographic examination. Differentiating a hypertrophied

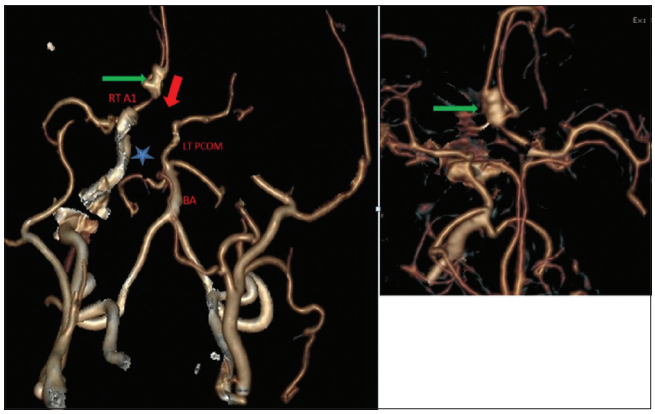

Figure 4: Three-dimensional reconstruction images show saccular aneurysm of the anterior communicating artery (green arrows), single left hypertrophied posterior communicating artery. Note the absence of right posterior communicating artery and A1 segment of the left anterior cerebral artery

PCOM from a persistent trigeminal artery are generally easily accomplished with either MR or CT angiography as the PCOM's origin from the supraclinoid ICA can be distinguished from the persistent trigeminal artery's origin from the cavernous ICA.

Nonvisualization of an ICA on angiography and absent bony carotid canal in the base of the skull on CT is the imaging findings that must be included in the diagnosis of congenital absence of an ICA. In the absence of any of these findings, it may be confused with acquired stenosis or occlusion of the ICA, respectively.

Intracranial vascular anomalies can be associated with ICA agenesis as seen in our case. A higher incidence of intracranial aneurysm (25%–43%) has been reported in association with ICA agenesis in comparison to the general population (2%–4%).[11,7] An aneurysm may develop secondary to hemodynamic disturbances or may form during embryonic life as a result of the developmental anomaly. In a series of six patients, Lee *et al*. reported that aneurysms developed ipsilateral side to the absent ICA, supporting a congenital origin of an aneurysm as opposed to hemodynamic factors.[11] The saccular ACOM artery aneurysms in our case may have arisen secondary to hemodynamic stress on the right ACA and ACOM by supplying the left ACA.

Our patient has no complaints at present and is being followed up with periodic physical and neurological examination.

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Conflicts of interest

There are no conflicts of interest.

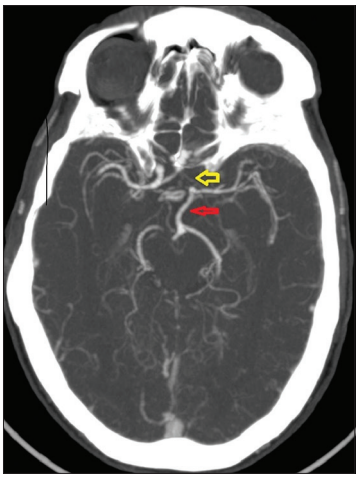

Figure 5: Computed tomography maximum intensity projection reconstruction image shows collateral supply to the left cerebral hemisphere through a saccular aneurysm of the anterior communicating artery and hypertrophied posterior communicating artery (red arrow). Note the absence of right posterior communicating artery (asterisk) and A1 segment of the left anterior cerebral artery (yellow arrow)

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