Agenesis of the internal carotid artery: a case report*

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Abstract The present paper reports a case of a 14-year-old-female adolescent who presented a single episode of syncope, without any other symptom. Axial and coronal T2-weighted magnetic resonance imaging demonstrated an absent right internal carotid artery flow void. A subsequent magnetic resonance angiography utilizing the time-of-flight technique showed absence of the right internal carotid artery. This finding was confirmed by magnetic resonance angiography of the cervical vessels, and axial computed tomography angiography showed agenesis of the right carotid canal. The literature reports such finding in association with other anomalies such as transphenoidal encephaloceles and circle of Willis aneurysms. These associations were not observed in the present case. The patient remained asymptomatic. Keywords: Internal carotid artery; Agenesis; Syncope.

INTRODUCTION

The embryonal development of the cerebral arterial circulation occurs in seven phases. Certain alterations in this process may lead to agenesis or hypoplasia of the carotid vessels. According to Streeten(1), Padget(2), and McLone & Naidich(3) two branches of the primitive internal carotid artery (ICA) develop early in the embryogenesis, originating from the third aortic arch. In its primitive form, the ICS reaches the cephalic region up to the level of the Rathke’s pouch where two primary divisions will occur. Only one cranial branch will extend anteriorly to supply the developing forebrain. In summary, the anterior choroidal, middle cerebral, anterior cerebral and primitive olfactory arteries will develop from this vessel. Posteriorly, another branch will give rise to the posterior choroidal, diencephalic and mesencephalic arteries. As these branches advance caudally, anastomosis will be made with the developing longitudinal neural arteries supplied by the trigeminal artery connections to the primitive ICA.

Ultimately, this pattern describes the first and second developmental phases and results in the adult cerebral circulation model to be established after the alterations along the other five phases as follows: the third phase, involving the vertebral arteries; the fourth phase, involving the anterior cerebral artery; the fifth phase, ophthalmic arteries; the sixth phase, the circle of Willis; and the seventh or fetal phase(4).

CASE REPORT

A female, caucasian, 14-year-old patient, coming from the interior of the state of São Paulo, who, 15 days ago, had undergone an episode of syncope witnessed by her relatives, where transitory signs of somnolence and dysarthria were observed. The patient was immediately referred to a neurologist.

Both during the clinical examination and magnetic resonance imaging (MRI) scanning, the patient presented asymptomatic, with no new episode of syncope or other complaints being reported. The suspicion of ICA agenesis was raised by MRI...
in the absence of the typical flow void in the ICA region, on the spin-echo sequences (Figure 1).

Considering the suspicion of agenesis or hypoplasia of the right ICA, the patient was submitted to non-contrast enhanced computed tomography (CT) and MRI angiography of the brain and cervical vessels, for diagnostic confirmation.

The CT study was aimed at differentiating a congenital from an acquired agenesis of the bony carotid canal, but the absence of the right carotid canal was observed.

The MRI angiography of the brain and cervical vessels was performed in an attempt to depict a global picture of the cerebral arterial network, and the absence of the right ICA from the aortic arch (Figures 2, 3 and 4).

Additionally, the patient was submitted to multislice CT, in an attempt to demonstrate some other skull bone abnormality, but only a confirmation of the other studies findings was observed by means of the 3D imaging reconstruction with volume rendering, demonstrating the absence of the right carotid canal.

**DISCUSSION**

An extremely rare incidence of ICA agenesis has been reported, with few scarce tens of cases being worldwide reported, maybe, to some extent, because the dysgenesis is generally asymptomatic in the greatest majority of patients. This occurs because there is a sufficient cerebral circulation supplied by anastomosis in the circle of Willis, intracavernous and external carotid artery anastomosis, besides persistent embryonal arteries. In these cases, the patients are referred for medical assistance because of complications resulting from abnormalities associated with carotid artery agenesis.

The ICA agenesis is usually unilateral, although there are reports of asymptomatic bilateral agenesis. If the agenesis is detected by MRI angiography, it must be confirmed by CT, in an attempt to find hypoplasia or absence of the carotid canal. Generally, the main secondary source of blood supply is the vertebrobasilar system (in cases of bilateral agenesis) or the dominant ICA (in cases of unilateral agenesis or hypoplasia).

The main findings associated with these anomalies are transsphenoidal encephaloceles, circle of Willis aneurysms, and an extensive rete mirabilis in the cranial base. Intracranial aneurysms are found in about 25% of cases of symptomatic internal carotid artery agenesis with intracranial hemorrhagic manifestations.

Also, other less frequent findings are reported. There are few cases where agenesis is associated with neuropsychomotor development delay, agenesis of the corpus callosum and persistent cavum vergae, in patients with bilateral agenesis. Few cases of unilateral agenesis in association with arachnoid cyst are reported. Also, there is a report about association of megadolicho-choanal anomaly and olivopontocerebellar atrophy with unilateral ICA agenesis, besides rare cases of hypopituitarism associated with unilateral artery agenesis, although intracavernous anastomosis seem to be efficient.

The absence of clinical or neurological symptoms of ischemia in the majority of patients with unilateral or bilateral agenesis or hypoplasia of the ICA leads to the assumption that, most of times, perfusional

![Figure 1](image1.png) Initial coronal MRI turbo spin-echo T2-weighted sequence showing absence of flow void in the right ICA.

![Figure 2](image2.png) Non-contrast-enhanced, axial CT, bony window technique showing the absence of the right carotid canal.
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to confirm the patency or dysgenesis of the carotid canal, corresponding to the vessel agenesis or hypoplasia\(^{16}\). Multidetector CT with 3D reconstruction also is useful in cases of dubious diagnosis.

**CONCLUSION**

In this brief case report, there is no intention to affirm that agenesis of the ICA should be always suspected. However, it may be concluded that a detailed and methodological evaluation of the carotid canals and ICA flow void in the investigation of primary or acquired stenosis (a usual finding in patients with neurological complaints) may lead to the finding of this anomaly that, although asymptomatic, may be associated with other potentially severe malformations and disorders.

The carotid canals should always be evaluated during the reading of routine cranial CT studies with axial slices and bone window images, considering that this is the confirmatory sign of the ICA absence, and does not imply an increase neither in the acquisition time nor in the cost of the study.

Likewise the present case, other cases of agenesis of the internal carotid artery have been incidentally found by MRI angiography. In these cases, it is always necessary to confirm if the hypoflow is caused by an acquired stenosis or dysgenesis of the vessel and the bony carotid canal. So, axial CT of the skull base should be performed to confirm the patency or dysgenesis of the carotid canal, corresponding to the vessel agenesis or hypoplasia\(^{16}\). Multidetector CT with 3D reconstruction also is useful in cases of dubious diagnosis.

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