CASE REPORTS

# RIGHT AORTIC ARCH ASSOCIATED WITH AGENESIS OF THE LEFT INTERNAL CAROTID ARTERY – AN EXTREMELY RARE CASE REPORT

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## Abstract

***Introduction:*** *A right aortic arch and agenesis of internal carotid artery (ICA) are both extremely rare vascular development anomalies. Etiology of the both anomalies might be associated with the abnormal regression of the dorsal aorta. Most cases of ICA are asymptomatic due to sufficient collateral circulation and it is usually an incident finding on head and neck imaging by color Doppler ultrasonography, computed tomography (CT) or magnetic resonance imaging (MRI).*

*ICA agenesis has a significant association – 24-67% – with intracranial aneurysms and their early detection can spare the patient serious complications.*

***Clinical case:*** *A 28-year-old male had a single episode of hypertension that motivated the realization of several tests. During the investigation he was submitted to a duplex ultrasonography that revealed a diffuse narrowing of the left common carotid artery (CCA), with a markedly decrease in the peak systolic velocity and the absence of the left internal carotid artery (ICA) was suspected. Contrast-enhanced computed tomography (CT) demonstrated no abnormalities, such as cerebral infarction or intracranial vascular malformations, but confirmed a right-sided aortic-arch, with anomalous origin of the left subclavian artery with a common origin of both CCAs and the absence of the left ICA. Examination of the head CT in bone window demonstrated an absence of the left internal carotid canal.*

***Conclusion:*** *This clinical case emphasizes the importance of recognizing this condition due to the associated hemodynamic changes and in order to discover and evaluate other additional vascular malformations (aneurysms, collateral channels) and their life threatening potential risks (subarachnoid hemorrhage or ischemia). Also, it has a special importance in case of planning carotid or trans-sphenoidal hypophyseal surgery. To our knowledge, only 8 cases have been reported right aortic arch associated with agenesis of the left internal carotid artery.*

***Keywords:*** *Carotid artery; Agenesis; Intracranial aneurysm; Right aortic arch*

## INTRODUCTION

The internal carotid artery (ICA) agenesis is a rare malformation disorder. This condition is estimated to occur in less than 0.01% of the population.[1,2] Similarly, a right aortic arch is an unusual congenital anomaly with an incidence of 0.05–0.1%.[3] Agenesis of the internal carotid was described for the first time in 1787, post-mortem, and in vivo in 1954, after an angiography examination.[4]

The most frequent congenital anomalies of the ICA can be classified as agenesis, aplasia and hypoplasia, and may be unilateral or bilateral.[5]

From the hemodynamic point of view the lack of flow through the carotid system is compensated, in most

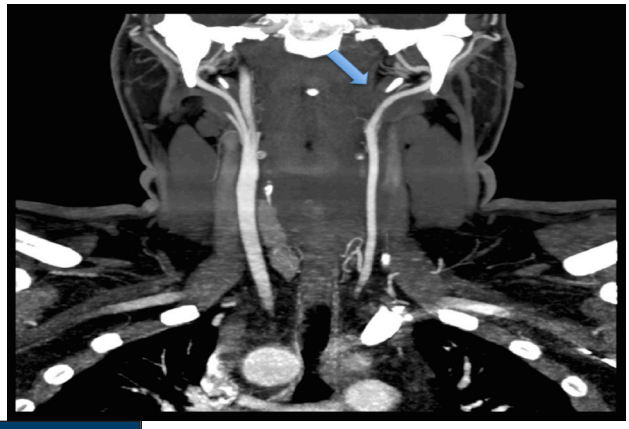

**Figure 1** *Computed tomographic angiography scan. Images show hypoplasia of the left common carotid artery with absence of left internal carotid artery (arrow).*

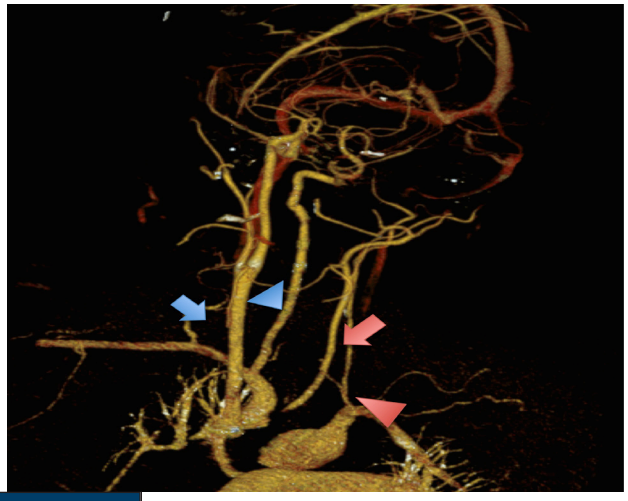

**Figure 2** *Reconstruction of computed tomographic angiography scan. Blue arrow, right common carotid artery (CCA); blue head arrow, right vertebral artery; red arrow, left CCA; red head arrow, left vertebral artery*

cases, through the circle of Willis and, less frequently, by the persistence of embryological vessels or by collaterals. However, when symptoms are present they include recurrent headache, blurring vision, hearing loss, hemiparesis with or without cranial nerve palsy, Horner syndrome, and intracranial hemorrhage due to ruptured aneurysms.6

We report the rare case of a right aortic arch associated with agenesis of the left internal carotid artery.

## CLINICAL CASE

A 28-year-old male had a single episode of hypertension with headaches. His physical and neurological examinations were normal and his medical history was unremarkable. During the investigation he was submitted to a duplex ultrasonography that revealed a diffuse narrowing of the left common carotid artery (CCA), with a markedly decrease in the peak systolic velocity and absence of the left internal carotid artery (ICA) was suspected.

Angio computed tomography (CT) demonstrated no abnormalities, such as cerebral infarction or vascular malformations, but confirmed a right-sided aortic arch, with anomalous origin of the left subclavian artery, a common origin of both CCAs and the absence of the left ICA (figure1, 2 and 3).

On careful review of the angio CT, the anterior communicating artery (ACOA) supplies the anterior cerebral artery (ACA) and the posterior communicating artery (PCOA) supplies the middle cerebral artery (MCA).

Examination of the head CT in bone window demonstrated an absence of the left internal

carotid canal (figure 4). The patient remains asymptomatic and attends regular follow-up consultations, which consists in an annual follow-up with color Doppler ultrasound and the monitoring of cardiovascular risk factors - smoking, hypertension, Diabetes and hypercholesterolemia.

## DISCUSSION

The cervical and cerebral arterial system experiences many transformations during the process of embryonic development before its final form in the fetus. Development of this system is modulated by numerous molecular factors and failures in these pathways can cause anatomic variants and a range of different clinical repercussions.

Early in the third gestational week, the cervical aortic primitives run in pairs as dorsal and ventral aortic roots. Six primitive aortic arches connect the dorsal and ventral aortic roots and 6 intersegmental arteries run laterally from the dorsal aortic roots. The ventral aortic roots cranial to the third aortic arch become the ventral pharyngeal artery. The ventral second aortic arch eventually regresses. The dorsal second aortic arch becomes the hyoid artery and stapedial artery. The stapedial artery connects the ventral pharyngeal artery and separates from the hyoid artery at the 24-mm embryonic stage (approximately the seventh gestational week). The external carotid artery (ECA) develops from the combination of the ventral pharyngeal artery and stapedial artery. The ICA derives from the third aortic arch and the

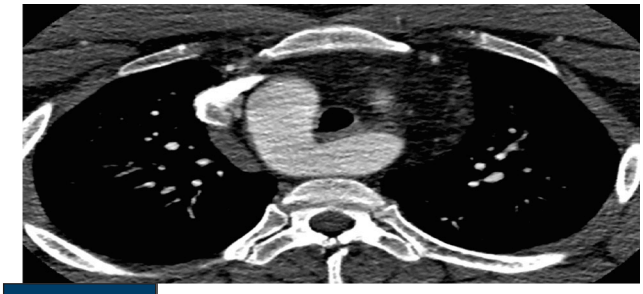

**Figure 3** *Computed tomographic angiography scan – axial plane. Right-sided aortic arch.*

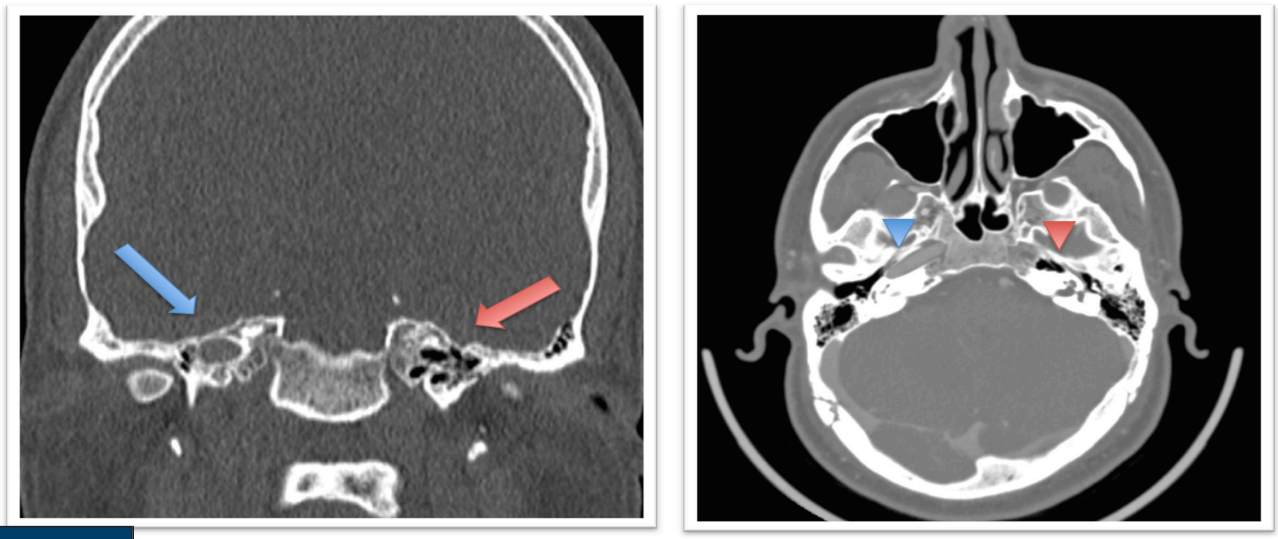

**Figure 4** *Computed tomographic angiography scan. Images show normal flow in the right intracranial internal carotid artery (ICA) (blue arrow and blue head of arrow) and no visibility of the left intracranial ICA and carotid canal (red arrow and red head of arrow), consistent with agenesis.*

dorsal aortic roots cranial to the third aortic arch. The CCA evolves from the fusion of the ventral pharyngeal artery and ventral third aortic arch. This development pattern occurs in approximately 65% of the population; other variants are observed in the remainder. The carotid canal formation occurs at the same time of this process and it is dependent on the development of the ICA, therefore, the congenital absence of the artery is accompanied by the absence of the carotid canal.[7,8]

The left fourth arch and dorsal aorta form the normal left aortic arch. Regression of the right dorsal aortic root (between the right subclavian artery and descending aorta) leaves the normal left aortic arch. However, in cases with right aortic arch, the right dorsal aortic root persists and a right aortic arch is formed by regression of the left fourth arch and left dorsal aorta during embryonic development.[7,8]

If there is a carotid malformation, the collateral blood flow pattern and the intracranial

vasculature depend on the stage at which development of the artery is interrupted. Cali et al.[9] suggested that if development of the internal carotid artery is interrupted before the conclusion of the circle of Willis, then collateral circulation would occur through the primitive vessels (intercavernous anastomoses). If the interruption occurs after the circle is concluded, then collateral circulation will predominantly flow through this route.

The development anomalies of the ICA can be classified as agenesis (complete failure of arterial development – absence of both ICA and carotid canal), aplasia (lack of development – presence of vestiges of non-patent vessels and also of the carotid canal), or hypoplasia (incomplete development of the artery – one of the ICA with a reduced but patent vascular lumen accompanied by a smaller but normally structured carotid canal).[4,10] Regarding the absence/presence of the bony carotid canal, it can be observed on the CT scan of the skull base. This information has a particular importance since it will differentiate aplasia from agenesis/hypoplasia (associated with a diminutive carotid canal)[5,7] – in our clinical case, examination of the head CT in bone window demonstrated an absence of the left internal carotid canal, findings that categorizes our case as a carotid agenesis, rather than hypoplasia or aplasia.

The causes of agenesis/aplasia of the ICA could be mechanical (pressure effects), excessive bending of the cephalic end of the embryo to one side or the other or constrictions by amniotic bands.[11]

Lie et al[12] described six collateral circulation patterns associated with absence of ICA (figure 5): Type A – collateral circulation to the ipsilateral ACA through a patent ACOA and to the ipsilateral MCA from the posterior circulation through a hypertrophied PCOA; Type B – the ipsilateral ACA and MCA are supplied across a patent enlarged ACOA; Type C – bilateral agenesis of the ICA with supply to the anterior circulation via carotid-vertebro-basilar anastomoses through hypertrophied PCOAs; Type D – unilateral agenesis of the cervical portions of the ICA with an inter-cavernous communication to the ipsilateral carotid siphon from the contralateral ICA; Type E – the ACAs are supplied by bilateral hypoplasic ICAs and the MCAs are supplied by enlarged PCOAs; Type F – collateral flow provided via transcranial anastomoses from the homolateral internal maxillary branches of the ECA system (rete mirabilis).

A simplified classification of collateral pathways can summarize the six types mentioned above into three main types as follows: the fetal type – the ACOA supplies the ACA, and the PCOA supplies the MCA (the one present in the case we described) – the most common type;

the adult form, where the ACOA supplies both ACA and MCA; the third type in which posterior collateral path-

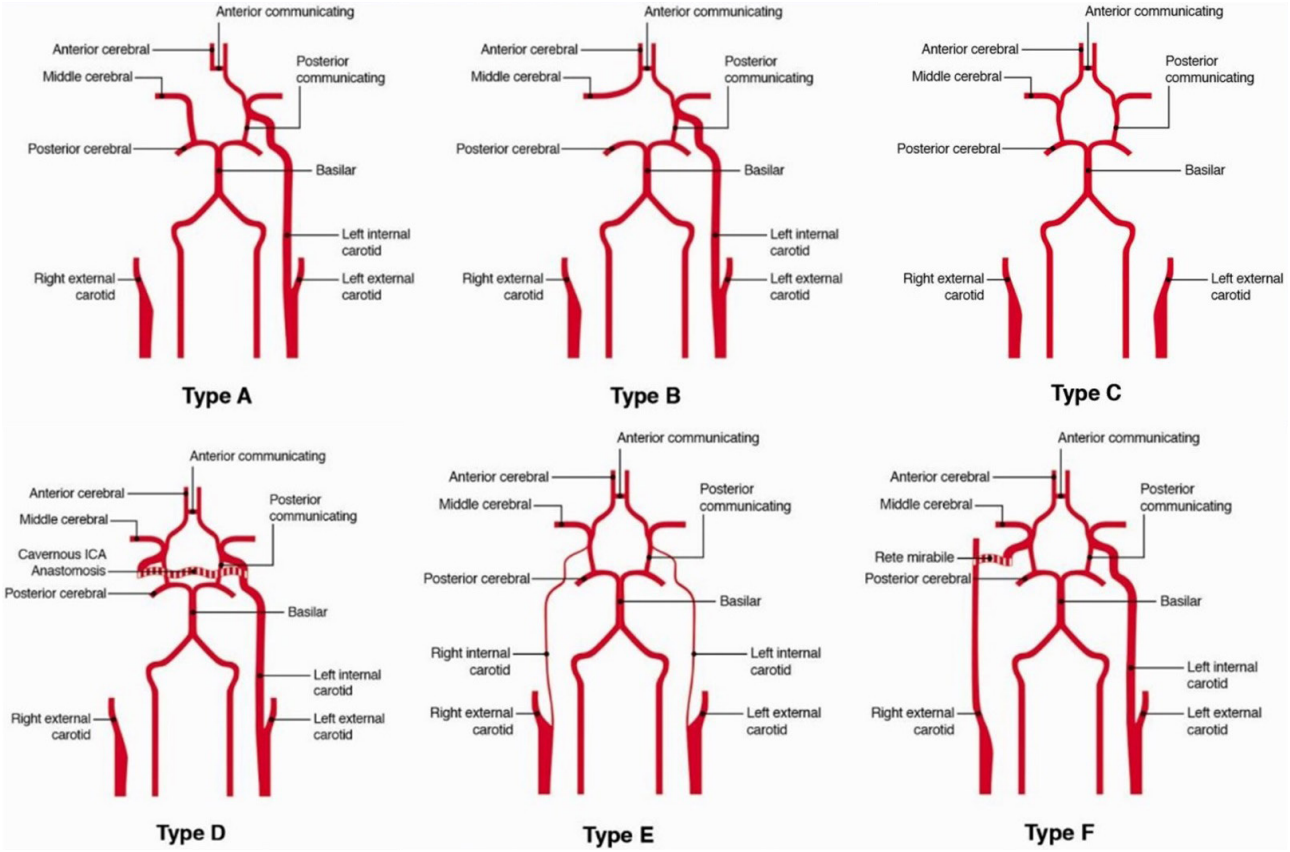


**Figure 5** *Collateral circulation patterns associated with absence of internal carotid artery (ICA). Illustration of different types of anastomosis in the setting of ICA agenesis described by Lie et al.*

ways through anastomosis from the ECA, contralateral ICA or from some primitive vessels (the rarest).[4,7]

Agenesis of the internal carotid is generally unilateral. In these cases, the principal blood supply compensating for the absence is the contralateral internal carotid, as mentioned above. A slight predominance among men and a preference for the left, at a ratio of 3:1, have been reported.[13]

Since this anomaly is asymptomatic in the majority of cases, it is generally diagnosed as an incidental finding in imaging exams. However, an association with some anomalies in the central nervous system, such as cerebral hemiatrophy, arachnoid cyst, neurofibromatosis, and Klippel-Feil syndrome, has been reported.[12] The most usual pathology related to ICA agenesis is intracerebral aneurysms with a reported incidence of 24-67%[13], which is much higher than that found in the general population (2–4%).[14] Increased flow through collateral vessels and altered flow dynamics are responsible for this increased prevalence, being the anterior communicating artery the most frequent site of aneurysm formation in such cases.[4]

Magnetic resonance angiography (MRA) or CT is recommended in the initial evaluation of ICA agenesis to evaluate for intracranial saccular aneurysm, particularly after the third decade of life. In the case we herein described, there were no detectable intracranial aneurysms.

## CONCLUSION

To our knowledge, only 8 cases of right aortic arch associated with agenesis of the left internal carotid artery have been reported in literature.[3, 15-21]

This clinical case emphasizes the importance of recognizing this condition due to the associated hemodynamic changes and in order to discover and evaluate other additional vascular malformations (aneurysms, collateral channels) and their life threatening potential risks (subarachnoid hemorrhage or ischemia). No guidelines currently exist pertaining to the management of carotid stenosis with contralateral agenesis, and therefore indications for treatment are the same as in the general population, which highlights the necessity of a special importance in case of planning carotid or trans-sphenoidal hypophyseal surgery.

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