CASE STUDY

Unilateral Agenesis of the Internal Carotid Artery

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Abstract Unilateral agenesis of the internal carotid artery (ICA) is an extremely rare anomaly. Diagnosis is often incidental in a radiological examination. Recognition of this anomaly has important clinical implications because other life-threatening conditions can be associated and it must be taken into account when planning carotid surgery. We report a case of 64-year-old man with agenesis of the ICA.

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Introduction

Agenesis of the internal carotid artery (ICA) is a rare congenital anomaly that was first described in 1787 by Lie. Since then, nearly 150 cases of congenital anomalies of the ICA have been described in the medical literature, ranging from its complete absence to its aplasia and hypoplasia.

In most cases, patients remain asymptomatic for long periods of time due either to the development of compensatory collateral circulation via the circle of Willis, or to the persistence of embryonic arteries, which are sufficient to maintain cerebral perfusion. Diagnosis usually takes place incidentally, during a radiological examination performed for other reasons.

We present a case of agenesis of the ICA for the purpose of raising awareness of this rare entity, as well as its possible clinical and surgical implications.

Clinical Case

In November 2009, a 64-year-old male was examined at our service due to left ear hearing loss of 1 month's
Figure 1  Axial CT scan without contrast of the petrosals, showing the absence of the left carotid canal at the base of the skull. ☆: Right carotid canal; *: absence of left carotid canal.

The spectrum of this malformation is variable and can range from complete absence of the artery, as in our patient, to the absence of a single section.

The cause of this malformation is unknown. The ICA stems from the third aortic arch and its formation is essential for further development of the carotid canal. Therefore, in our case, given the absence of the carotid canal, the alteration would have occurred between the third and fifth week of embryonic development, during which the ICA is defined, prior to the formation of the carotid canal at the base of the skull. Furthermore, although our patient presented slight hypoplasia of the CCA, the external carotid artery (ECA) was normal. Consequently, a different embryonic origin of the internal and external CAs is likely.

The ICA gives rise to the anterior and middle cerebral arteries, which irrigate most of the cerebral territory. The lack of flow through the carotid system is compensated, in most cases, through the circle of Willis and, less frequently, by the persistence of embryological vessels or by collaterals. Three alternative circulation patterns have been described. There was a "foetal type" in our case, in which the anterior cerebral artery (ACA) on the affected side arises from the contralateral anterior communicating artery and the middle cerebral artery (MCA) begins at the basilar system via a hypertrophic posterior communicating artery or a persistent trigeminal artery. In the "adult type", the ACA and MCA originate in the anterior communicating artery and, in the third type (the rarest), there is anastomosis between the external and internal carotid systems.

Agenesis or aplasia of the ICA is normally asymptomatic due to the development of substitutions and its diagnosis is coincidental, as in the present case. These patients may consult due to symptoms such as repeated headaches, hearing loss, vision alterations, paresis of cranial nerves or even Bernard–Horner syndrome. The diagnostic study of these patients involves imaging studies and agenesis of the ICA is diagnosed, with no causal relationship to the symptoms being found. However, associated anomalies such as cerebral aneurysms or abnormal vascular communications should alert about the possibility of subarachnoid haemorrhage or ischemic phenomena, if the collateral circulation is insufficient. The incidence of aneurysms of the circle of Willis, mainly in the anterior communicating artery, can reach 34%, a higher figure than that found in the general population (2%-4%). This could be due to an increase in the flow of the supplementary arteries (flow aneurysms). Other, less frequent, associated abnormalities are type II neurofibromatosis, coarctation of the aorta, polycystic kidney disease, Klippel–Trenaunay syndrome and agenesis of the corpus callosum.

This anomaly is diagnosed by CT angiography or ultrasound angiography, and CT demonstrates the absence of a carotid canal. It should be noted that having a single carotid axis may lead to cerebrovascular events on the affected side, due to contralateral atheroma or even bilateral ischemic lesions. Furthermore, it is essential to be aware of the absence of an ICA when performing interventional manoeuvres or carotid surgery on the healthy side.

Discussion

Agenesis of the ICA is a rare, congenital anomaly. Its true incidence is unknown because it is generally a coincidental finding during a radiographic examination for other reasons. Affifi et al. estimate a real frequency below 0.01% of the general population.

As in the case presented here, ICA agenesis is more common on the left side but may also be bilateral. In addition, the physical examination found no data of interest and otoscopy was consistent with normality. The audiometry performed revealed the presence of deafness in the left ear and normal hearing with correct verbal discrimination in the right ear.

Given these findings, we requested a computed tomography (CT) of the petrosals so as to exclude morphological abnormalities in the left petrosal. This did not identify fractures, dislocations of the ossicular chain or any post-traumatic sequelae that justified the loss of hearing. However, an absence of the left carotid canal was found incidentally, which translated into an ipsilateral ICA agenesis (Fig. 1). For this reason, the radiologist recommended performing a CT angiography of the supra-aortic trunks (SAT) and the circle of Willis, to rule out abnormalities associated with intracranial vasculature. This exploration confirmed the absence of the left ICA, as well as secondary hypoplasia of the ipsilateral common carotid artery (CCA). No associated vascular malformations were observed (Fig. 2). In addition, we observed that anterior cerebral circulation on the right side was completed via the anterior communicating artery and through a persistent trigeminal artery.
Conclusion

Agenesis of the ICA is an exceptional congenital anomaly whose diagnosis is often coincidental, since it causes no symptoms in the majority patients. Its interest lies in the fact that it is sometimes associated with malformations of the circle of Willis (such as aneurysms) and, therefore, these patients have a higher risk of cerebrovascular events. Its diagnosis and that of associated anomalies are made possible by CT and CT angiography.

Conflict of Interests

The authors have no conflicts of interest to declare.

References