Case Report

Computed tomography angiography of unilateral agenesis of the internal carotid artery: 2 cases report with focus on embryology, collateral pathways, and imaging

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\textbf{Abstract}

Congenital absence of internal carotid artery (ICA) is a rare anomaly seen in around 0.01% of the population. High incidence of aneurysms is reported in these patients population. Many patients are asymptomatic as the development of a collateral circulation ensures cerebral perfusion. The embryology, the common collateral pathways, and the imaging findings associated with this anomaly are illustrated with 2 new cases. We reported the cases of 2 totally asymptomatic patients at the time of imaging in which ICA agenesis was proved on computed tomography angiography. On imaging, all the most important findings necessary for ICA agenesis diagnosis have been identified and described. Noninvasive imaging techniques are currently the mainstay of ICA agenesis diagnosis.

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\textbf{Introduction}

The agenesis of the internal carotid artery (ICA) is characterized by the simultaneous absence of the ICA and the carotid canal at skull base, due to an embryologic arterial developmental failure. Although this anatomic anomaly has been described already in XVIII century (Tode first described a case with the absence of the ICA in 1787), nowadays, less than 100 cases have been reported in literature [1]. ICA agenesis is usually discovered incidentally by means of head-and-neck computed tomography (CT) or magnetic resonance imaging examinations. The absence of the ICA requires the existence

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of alternative pathways to ensure the ipsilateral vascularization. In the ICA agenesis, Lie described 6 pathways of collateral circulation, named as type A to type F [2,3]. Type A refers to the unilateral absence of the ICA and collateral circulation to the ipsilateral anterior cerebral artery (ACA) through an anterior communicating artery (ACOM) and to the ipsilateral middle cerebral artery (MCA) from the hypertrophic posterior communicating artery (PCOM). Type B refers to the unilateral absence of ICA and collateral flow to the ipsilateral ACA and MCA through a patent ACOM artery. Type C refers to the bilateral ICA agenesis with supply to the anterior circulation through hypertrophic PCOM. Type D refers to the unilateral agenesis of ICA with an intercavernous communication to the ipsilateral carotid siphon from cavernous segment of contralateral ICA. Last, type E refers to the bilateral hypoplastic ICAs supplying ACA and the MCAs are supplied by enlarged PCOMs, and type F refers to the flow to ICA across transcranial anastomosis from the external carotid artery (so-called “rete mirabile”).

We presented 2 patients with left ICA agenesis focusing our analysis on embryologic and radiological findings.

**Cases description**

**Case 1**

A 42-year-old female was admitted to our gynecology department for endometriosis surgery. Her neurologic examination was normal, but her previous medical history reveals some previous episodes of blurring vision. For this reason, the patient underwent a sonographic examination of epiaortic vessels which revealed a suspected occlusion of the left ICA. Subsequently, the patient was evaluated with CT angiography highlighting the most important findings of the ICA agenesis, that is, the absence of the carotid canal at the skull base, the absence of the ipsilateral ICA, and the hypoplasia of ipsilateral common carotid artery (Fig. 1). CT angiography study also showed an hypertrophy of vertebrobasilar circulation and a collateral pathways cerebral circulation. In particular, we observed that the ipsilateral ACA is supported by the ACOM and that the ipsilateral MCA is supported by the hypertrophic PCOM. According to the Lie classification [2], this alternative pathway is classified as type A.

**Case 2**

A 73-year-old man was admitted to our department of Cardiothoracic Surgery for a coronary artery disease. His neurologic examination was normal, and the patient had no previous significant medical history. A routine sonographic evaluation of the carotid arteries revealed a suspected left ICA agenesis. Subsequently, the patient was evaluated with CT angiography which demonstrated the absence of the carotid canal at the skull base, the absence of the ipsilateral ICA, and the hypoplasia of ipsilateral common carotid artery (Fig. 2). Moreover, CT angiography study also showed a collateral pathways cerebral circulation. In particular, we observed that a patent ACOM artery supports the ipsilateral ACA and MCA. According to the Lie classification [2], this alternative pathway is classified as type B.

Fig. 1 – Computed tomography (CT) volume rendering (VR) reconstructions show (A) the hypoplasia of ipsilateral common carotid artery (yellow arrowhead) and (B) the absence of the ipsilateral ICA (blue arrowhead). CT axial image at the level petrous ICA shows (C) the absence of the carotid canal to the skull base (yellow arrowhead). VR reconstructions of the circle of Willis show (D) the type A of collateral pathways cerebral circulation according to the Lie classification and an hypertrophy of vertebrobasilar circulation (red arrowhead). ICA, internal carotid artery.
CT examination also showed another patient’s congenital anomaly, the corpus callosum agenesis.

Discussion

Congenital absence (agenesis) of ICA is a rare vascular disorder incidentally discovered by imaging and whose incidence is about 0.01% [4]. Agenesis is a complete developmental interruption of ICA with the absence of the ipsilateral carotid canal; more frequently this anomaly involves the left ICA [5]. The absence of the bony carotid canal is essential to differentiate this anomaly from chronic ICA occlusion.

The embryology knowledge allows us to understand the anatomic variants. During human embryo development, 6 pairs of arteries (the so-called aortic arches) are contained in the branchial apparatus mesenchymal tissue, running through the pharyngeal arches.

The aortic arches originate from the most distal part of the truncus arteriosus (the aortic sac), pass through pharyngeal arches, and terminate in the right and left dorsal aortae.

The third arches are precursors to the carotid system. The carotid system develops after the involution of some arterial segments. After 29 days, the first and second arches regress; subsequently, within the sixth week, also the carotid duct (a dorsal aorta segment extending between the third and fourth arcs) regresses. The regression of these arterial segments determines the persistence of a predominant channel of blood flow to the cephalic region, starting from the ventral aortic sac through the third aortic arch until the cranial extension of the dorsal aorta. The combination of these 3 arterial segments originates the common and internal carotid arteries. The external carotid artery is a bud of the third aortic arch; at 40 days, the migration of the external carotid origin determines the site of the carotid bifurcation [6]. ICA is completely formed after 6 weeks of fetal life [5]. Agenesis of ICA results from atresia or involution of the third aortic arches and the distal portion of the dorsal aortas, in the 20- to 24-mm stage [7].

Many patients are asymptomatic, as the development of a collateral circulation ensures cerebral perfusion.

Possible symptoms include recurrent headache, hearing loss, hemiparesis, blurring vision, ipsilateral Horner’s syndrome, and intracranial hemorrhage due to ruptured aneurysms. ICA agenesis is associated with 25%-34% of cerebral aneurysm formation [5]; increased flow through collateral artery is the possible cause of this increased prevalence.

Other abnormalities such as corpus callosum agenesis, meningocele, aortic coarctation, neurofibromatosis, and cardiac anomalies may be associated with agenesis of the ICA [8].

A robust diagnosis of agenesis of the ICA using CT and magnetic resonance imaging must prove the absence of the carotid canal to the skull base, the absence of the ipsilateral ICA, and the hypoplasia of ipsilateral common carotid artery. From an embryologic point of view, the ICA development occurs only after the formation of the carotid canal, so that the absence of the carotid canal generates an ICA agenesis [9].

In conclusion, our 2 cases have stressed all the important imaging findings that are required for the correct diagnosis of ICA agenesis rather than ICA occlusion. In fact, in the ICA occlusion, a normal carotid canal is always present and
characteristically the common carotid artery is not hypoplastic.

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