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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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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.

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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Informed consent: Informed consent was obtained from all individual participants included in the study.
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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.

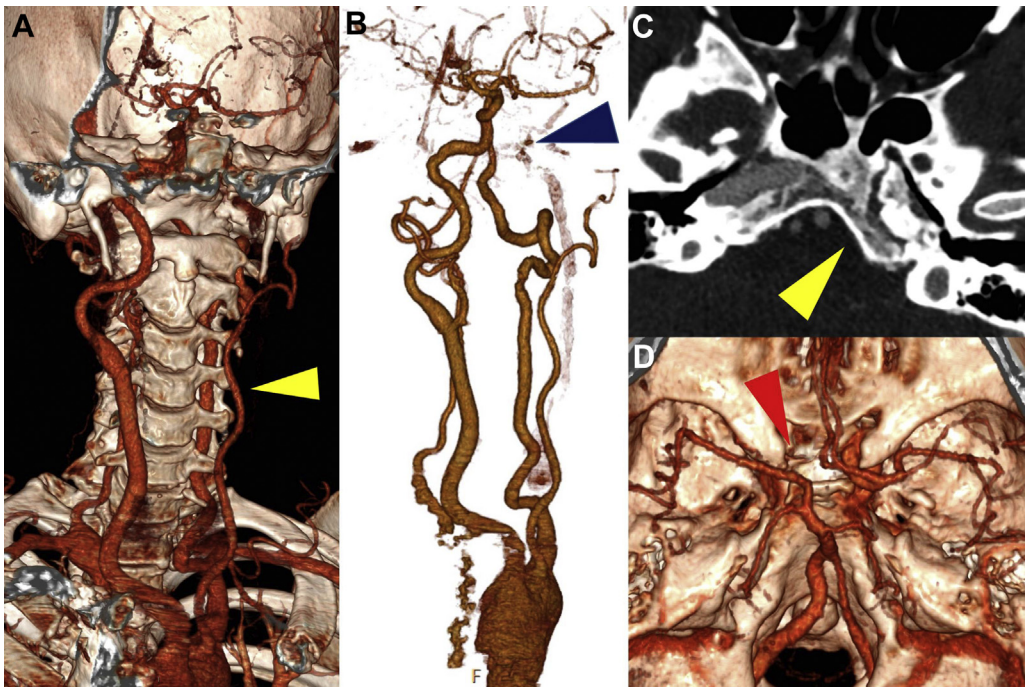

Fig. 1 – Computed tomography (CT) volume rendering (VR) reconstructions show (A) the hypoplasia of ipsilateral common carotid artery (yellow arrowhead) and (B) the absence of the ipsilateral ICA (blue arrowhead). CT axial image at the level petrous ICA shows (C) the absence of the carotid canal to the 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.