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Bilateral internal carotid agenesis: value of CT angiography and correlation to embryogenesis

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Abstract Bilateral internal carotid artery agenesis is an uncommon disease, difficult to differentiate from bilateral carotid artery thrombosis. A few case reports have described the contribution of conventional angiogram to make the diagnosis and recognize the anatomic details of this rare malformation. Advantages of CT angiography as a non-invasive radiologic tool are discussed in this case report.

Key words Carotid · Agenesis · CT angiography

Introduction

Bilateral internal carotid artery (ICA) agenesis is rare. A few cases have been reported in the literature [1]. This anomaly is associated with skull base and vascular abnormalities. In a short series of abnormal development of ICA, including one case of bilateral ICA agenesis, Quint et al. emphasized the importance of scrutinizing the skull base on routine CT examination to detect asymmetric or absence of carotid canals [1]. In such cases, contrast-enhanced CT should be performed to look for underlying vascular abnormalities. We illustrate by this additional case of bilateral ICA agenesis the value of CT angiography in identifying skull base abnormalities and presence of patent a primitive trigeminal artery.

Case report

An 18-year-old man presented since childhood learning difficulties and neurologic symptoms including headaches, cortical blindness and epilepsy. Physical examination revealed an asymmetric eye size. Helical CT angiography with millimetric section thickness reconstruction on the skull base after intravenous contrast administration was performed and showed absence of carotid canal in the petrous pyramids (Fig. 1), absence of ICAs, and an important intra- and extra-cranial collateral vascular network. In this vascular network, we demonstrate important arterial supply from branches of the external carotid system (Figs. 2, 3) and the persistent of embryogenic artery by the primitive trigeminal artery (Fig. 4).

Discussion

Absence of ICAs development during the embryonic primordium results in absence of carotid canal and de-



Fig.1 Computed tomographic scan of the temporal bones demonstrates the absence of the carotid canals (*arrowheads*)



Fig.3 Same as Fig.2



Fig.2 Cervical portion CT angiography shows the absence of internal carotid arteries (*white arrowheads*) and asymmetric collaterals through the external carotid arteries supplying the internal carotid territories (*black arrows*)

development of a collateral circulation [1]. In our case, CT angiography findings provided information on both skull base and vascular changes which are well correlated to embryology. According to Streeter [2] and Padget [3], the skull base begins to take shape between the fifth and sixth weeks of fetal life, 2 weeks after ICA formation. Therefore, failure of ICAS development results in absence of carotid canal on the skull base [4, 5, 6]. Absence of carotid canals on CT excludes the diagnosis of ICAS thrombosis. In our case, the absence of ICAs resulted in the development of a large vascular collaterality issued from the branches of the external carotid arteries (ECAs) and resulted in the persistence of embryonic arteries. Furthermore, CT angiography shows anastomoses between the carotid and the vertebro-basilar system via a patent primitive trigeminal artery (PTA). In utero, PTA supplies the basilar territory by the ICA before the development of the posterior communicating and vertebral arteries [7]. In bilateral ICA agenesis, the PTA does not regress and supplies retrogradely the carotid system [8]. Primitive trigeminal artery arises from the proximal cavernous internal carotid artery either by a lateral way or by a midline course to join the basilar artery [9]. The incidence of a persistent PTA in different series published in the literature ranges from 0.1 to 0.6% and is associated with different neurologic entities [5, 10, 11]. Although helical CT angiography is a not reliable technique in assessing the intracranial portion of the ICA, it provides enough information to confirm the diagnosis of bilateral internal carotid agenesis.



Fig. 4 Basilar trunk supplies the internal carotid territories by the primitive trigeminal artery (*arrowhead*)

Conclusion

Computed tomography angiography is an effective and non-invasive tool for the diagnosis and follow-up of patients with bilateral ICA agenesis. It demonstrates the absence of carotid canals, the collateral arterial network with the persistence of embryonic arteries as primitive trigeminal artery.

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