Internal carotid artery agenesis with stenosed intercavernous anastomosis: a case report

Hongzhou Duan¹, Liang Li¹✉, Guiping Zhao², Yang Zhang¹, Jiayong Zhang¹

¹Department of Neurosurgery; ²Neurology, Peking University First Hospital, Beijing 100034, China.

Abstract

We report a rare case of internal carotid artery agenesis with stenosed intercavernous anastomosis. A 59-year-old male patient presented with a new infarction in the left basal ganglia. Magnetic resonance angiography and cerebral angiography showed that the right internal carotid artery disappeared from the origin to the foramen lacerum segment, and there was an anastomotic artery with severe stenosis passed through the floor of the sella and in front of the cavernous sinus. The right A1 segment of the anterior cerebral artery was absent and A2 segment was supplied by the normal contralateral internal carotid artery via the anterior communicating artery.

Keywords: internal carotid artery, artery agenesis, cavernous sinus

Introduction

Occlusion of the internal carotid artery (ICA) due to atherosclerosis is common, but ICA agenesis is rare, in which the blood supply of the ipsilateral brain is always compensated by the contralateral ICA and vertebrobasilar system via the circle of Willis. It is very rare that the affected hemisphere is supplied by an intercavernous anastomotic vessel connecting bilateral ICAs. We report a case of ICA agenesis with intercavernous anastomosis.

Case report

A 59-year-old man complained of fatigue and dysphasia for one week, but without numbness, bucking, dysphagia and dizziness. Magnetic resonance imaging (MRI) examination showed a new infarction in the left corona radiate and basal ganglia region (Fig. 1A). There were also some small old infarctions in the bilateral cerebral hemisphere and the right cerebellum. Doppler ultrasonography showed an atherosclerotic plaque about 18.3 mm × 3.2 mm (length × width) in the left common carotid artery bifurcation (Fig. 1B), while it was difficult to find the right common carotid artery bifurcation at the same level as that of the left side although there was a similar spectral waveform. The patient was admitted to the hospital, and a magnetic resonance angiography (MRA) examination showed that the right common carotid artery continued to the external carotid artery (ECA) directly, and there was an intercavernous anastomotic vessel with stenosis connected bilateral ICAs which passed through the floor of the sella and in front of the cavernous sinus (Fig. 1D).

Further cerebral angiography was performed and showed that the right common carotid artery continued to the external carotid artery (ECA) directly, and there was...
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Fig. 1 Ultrasound, MRI and MRA images in a 59-year old man with internal carotid artery agenesis with stenosed intercavernous anastomosis. A: A new infarction in the left basal ganglia region. B: An atherosclerotic plaque (arrow) is located in the bifurcation of the left CCA. C: Absent carotid canal in the right petrosal bone (arrow). D: An intracavernous anastomotic vessel with regional stenosis (arrow) connects bilateral internal carotid artery and is located at the bottom of the sella.

Fig. 2 Cerebral angiography images of the anastomotic vessel and compensatory circulation. A: Right common carotid artery angiography shows the absence of the right internal carotid artery without stub (arrow). B: Right common carotid artery angiography in the late arterial stage, ophthalmic artery appears retrogradely (arrow), and right internal carotid artery and middle cerebral artery appears. C: Right posterior communicating artery is patent (arrow). D: 3D reconstructed image shows an intracavernous anastomotic vessel with severe stenosis (arrow) connecting bilateral internal carotid artery, A1 segment of the right anterior cerebral artery disappears, and A2 segment is compensated by contralateral side.
no stub at the beginning of the ICA (Fig. 2A). There were some collateral arteries connecting the right ECA and right ophthalmic artery, and the right ICA is also shown by the reversal flow of the ophthalmic artery (Fig. 2B). The right posterior communicating artery was open and the right middle cerebral artery appeared during vertebral angiography (Fig. 2C). Left ICA angiography showed that an intercavernous anastomotic vessel with severe stenosis connected bilateral ICAs, and A1 segment of the right anterior cerebral artery (ACA) was not shown. Right A2 segment was supplied by contralateral ACA via the anterior communicating artery (Fig. 2D). Fresh infarction of the left basal ganglion was suspected to be related to atherosclerosis of small perforating branches, and after conservative therapy with antiplatelet drugs, the patient recovered well and discharged one week later.

Discussion

ICA agenesis has been rarely reported and the pathogenesis is still unclear. Some researchers speculate that this might occur before the 24-mm stage of growth when the circle of Willis becomes embryologically complete[1]. Some others suspected that unilateral agenesis may be due to some mechanical and hemodynamic stresses on the embryo like exaggerated folding of the embryo towards one side and constriction by amniotic bands[2]. Several blood flow compensatory mechanisms may exist after unilateral ICA agenesis[1]. The first type is also the most common type, in which ACA of the affected side is supplied by the normal contralateral ACA via the anterior communicating artery and middle cerebral artery arises from the basilar artery through an enlarged posterior communicating artery. The second compensatory mechanism is the same as atherosclerotic occlusion of the ICA in adults, ipsilateral ACA and middle cerebral artery are both supplied by the normal contralateral ICA via the anterior communicating artery. The third type which is similar to our patient is extremely rare, the affected ICA is absent from the origin, and an intercavernous anastomotic vessel communicates bilateral ICAs. Only 19 cases have been reported till now[2-3].

The source of the anastomotic vessel was not clear; some researchers believed that it might be the fusion of bilateral primitive trigeminal arteries which failed to develop their normal connection to the basilar artery[1]. Some authors speculated that it may originate from the primitive maxillary artery or preexisting medial rami from the cavernous ICAs[2,4-6]. In our case, MRA and rotational cerebral angiography showed that the anastomotic vessel originated from the horizontal segment of the ICA in the cavernous sinus, and passed through the floor of the sella and in front of the cavernous sinus, connecting the contralateral ICA. The route that the anastomotic vessel passed indicated that it might not be originated from primary trigeminal artery or primary maxillary artery, because the primary trigeminal artery always originated from the posterior wall of the ICA, it went posterioriomedially and was connected to the posterior cerebral artery, and the primary maxillary artery originated from the lateral wall of the ICA and went forward and outward, then be connected to the maxillary artery. Some researchers found that plexiform channels around Rathke’s pouch connected bilateral ICAs in a 4- to 5-mm embryo research[1]. Staples reported a similar case and suspected that the anastomotic vessel might be inferior hypophyseal or capsular arteries[7]. Based on some anatomical researchs, Harris found some small branches inside the cavernous sinus segment of the ICA occasionally, which were called McConnell capsular artery connected bilateral ICA and provided the blood supply for the pituitary[8]. We believe that the plexiform channels around Rathke’s pouch, the hypophyseal artery reported by Staples, the small branches by Harris and the vessel we report here are the same vessel called McConnell capsular artery.

Approximately 20 cases have been reported till now, including this case. The age ranges from 7 to 69 years. Males are a little more than females, and the incidence of right- and left-sided occurrences is equal[2]. Four (20%) of 20 patients had intracranial aneurysms, which exceeds the 2% to 4% natural incidence of intracranial aneurysms. This might be explained by deranged hemodynamic forces or developmental errors[2]. Therefore, it is necessary to follow up these patients with imaging examinations. Additionally, the A1 segment of the ACA on the agenetic side was not seen in angiography in 17 of 20 cases. Some researcher attributed this to hypoplasia[1]. In our case, we cannot find any strip-like image in the place of the right ACA on MRA. Therefore, we suppose that the A1 segment of the agenetic side disappeared angiographically might be due to agenesis, but not hypoplasia.

The golden criteria to diagnose this disease may be cerebral angiography, while MRA and MRI examination are also helpful in showing the location of anastomotic artery and other tissues around. Computed tomography (CT) is very useful in illustrating the absence of the carotid canal[9]. In our case, we can also show the absence of the carotid canal in MRI images. As the anastomotic vessel is located at the bottom of the sella, it may be mistaken for pituitary microadenoma, and flow void signal in MRI is helpful in differential diagnosis.

There is a severe stenosis in the anastomotic vessel in this case, to our knowledge, it may be probably the first
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Case report to show stenosis in such an anastomotic vessel. Fortunately, there are many compensatory blood supplies from the right posterior communicating artery and right ECA. We speculate that the stenosis might be due to atherosclerosis, and the right ECA provides collateral blood supply to the right intracranial ICA is secondary to the stenosis of anastomotic vessel, but not congenital. If the patient suffers from cerebral ischemia due to progressive stenosis or occlusion of the anastomotic vessel in future, EC-IC bypass might be a good choice.

Patient consent

The patient has consented to the submission of the case report to the journal.

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