IBRO Neuroscience Reports 14 (2023) 462–465

Contents lists available at ScienceDirect

IBRO Neuroscience Reports

journal homepage: www.sciencedirect.com/journal/IBRO-Neuroscience-Reports

Interventional reperfusion for acute embolism of the middle cerebral artery trunk with contralateral internal carotid artery congenital absence

Jun-wei Wang [a,1], Yu Zhang [b,1], Yong-tao Yang [a], Xin Li [a], Qun-ling Zhan [a], Ge Jin [a,*,2]

[a] Department of Neurology, The Fifth People's Hospital of Chongqing, No. 24 Renji Road, Nanan District, Chongqing 400062, China
[b] Department of Interventional Radiology, The Fifth People's Hospital of Chongqing, No. 24 Renji Road, Nanan District, Chongqing 400062, China

ARTICLE INFO

Keywords:
Middle cerebral artery
Embolism
Congenital absence of internal carotid artery
Interventional reperfusion

ABSTRACT

Internal carotid artery congenital absence with acute embolism of the middle cerebral artery trunk is very rare. A 65-year-old female with a history of hypertension and atrial fibrillation was admitted to the neurology department of our hospital. Computed tomography of the head and neck showed no carotid canal of the petrous portion of the temporal bone; digital subtraction angiography (DSA) showed no left internal carotid artery and the right middle cerebral artery trunk occlusion. These results suggested acute embolism of the middle cerebral artery trunk with contralateral internal carotid artery congenital absence. Mechanical thrombectomy was performed, which had a good outcome. This case showed the vascular anatomy features of ICA congenital absence with contralateral large vessel acute occlusion, and it is essential to promptly identify the vascular variation during the interventional procedure.

1. Introduction

The cerebral blood flow comes from the carotid artery system and the vertebrobasilar artery system. The large vessels that supply blood to the cerebral arch on the aortic arch can have congenital morphological abnormalities, such as anomalous origin, abnormalities of course, cerebral arterial fenestration and primitive persistent carotid-vertebrobasilar artery abnormalities. Internal carotid artery (ICA) congenital absence is very rare, with an incidence of approximately 0.01% (Lee et al., 2003). Tode first found congenital absence of the ICA at autopsy in 1787 (Midkiff et al., 1995), and Verbiest reported the first case found through cerebral angiography (Claros et al., 1999). There are few reports of congenital absence of the ICA in currently available literature, particularly with contralateral anterior circulation artery occlusion. Here, we report a female patient admitted to our hospital with left ICA congenital absence and right middle cerebral artery (MCA) M1 segment acute cardiogenic embolism. She had a good outcome after emergency transcatheter thrombus aspiration and reperfusion treatment.

2. Case report

A 65-year-old woman who presented with impaired speech and left limb weakness was admitted to our department when she was undergoing a health checkup in our hospital in October 2021. She was considered to have had an in-hospital stroke. She did not take oral anticoagulants or antihypertensive drugs regularly, although she had a history of atrial fibrillation and hypertension. The neurological physical examination revealed the following: right-handedness, sleepiness, slurred speech, right gaze preference, left facial weakness, 0/5 in the left upper extremity and 0/5 in the left lower extremity, uncooperative with coordination movement and sensory examination, negative meningeal irritation sign, and negative pathological reflex. The National Institutes of Health Stroke Scale (NIHSS) score was 18 points. The electrocardiogram showed atrial fibrillation. Noncontrast computed tomography (NCCT) showed no intracranial haemorrhage, and the Alberta Stroke Program early CT score (ASPECTS) was 10 points. There were no obvious abnormalities in routine blood tests, blood biochemical indices or clotting.

Intravenous thrombolysis with recombinant tissue plasminogen activator (rt-PA) was performed immediately according to her body

* Corresponding author.
E-mail address: wjwwysn@163.com (G. Jin).
[1] Both authors equally contributed to this study.
[2] ORCID: 0000-0002-2522-6271

https://doi.org/10.1016/j.ibneur.2023.04.007
Received 16 October 2022; Accepted 30 April 2023
Available online 18 May 2023

weight (0.9 mg/kg, total 56 mg). The onset-to-needle time (ONT) was 39 min.We decided to implement bridge endovascular interventional therapy with the consent of the family members of the patient. Multimodal imaging such as CT angiography (CTA), CT perfusion (CTP) andMagnetic Resonance Imaging(MRI) for brain was not performed because of the symptom onset within 4.5 h of anterior circulation stroke. We performed right femoral artery puncture under local anaesthesia with lidocaine and sedation with dexmedetomidine.The tip of a 70-cm-long 8-Fr sheath (Cook Medical, Bloomington, IN) was placed at the distal end of the right common carotid artery (CCA). Digital subtraction angiography (DSA) showed occlusion of the M1 segment of the right MCA, and the collateral circulation classification of the American Society of intervention and Therapeutic Neuroradiology/Society of interventional radiology (ASITN/SIR) was grade 1 (Fig. 1A). The AXS Catalyst™6 distal access catheter (Stryker, Business and Technology Model Farm Road Cork, IRELAND) was coaxially guided with Synchro-10 Neuro Guidewires (0.10 in × 200 cm, Stryker, Salt lake City, UT) and Trevor Pro 18 microcatheter (0.21 in × 150 cm, Stryker, Salt lake City, UT), and the tip of the distal access catheter was navigated directly to the thrombus and engaged with the thrombus under fluoroscopic guidance. Then, suction was applied directly with a 50 ml syringe once, after which DSA showed that the MCA had complete reperfusion, and the modified thrombolysis in cerebral infarction (mTICI) grade was 3 (Fig. 1B). During the operation, we also found that the left anterior cerebral artery (ACA) territory was supplied by the right anterior circulation through the anterior communicating artery (ACoA) (Fig. 1A and B) via DSA. In addition, the left CCA directly continued to be the external carotid artery (ECA), and the territory of the left MCA was supplied by the ipsilateral posterior circulation through the posterior communicating artery (PCoA). The CT showed no left carotid canal, and the contralateral morphology of the carotid canal was normal (Fig. 1 E - G).

Immediate CT after thrombectomy showed a small high-density area in the right lenticular nucleus (Fig. 1 C). The NIHSS score was 7 points 24 h after thrombectomy. Diffusion-weighted magnetic resonance imaging (MR-DWI) showed that there were some infarcted lesions that invaded the right frontal lobe, insular lobe and lenticular nucleus (Fig. 1 D). The patient was discharged on the 10th day after thrombectomy, when her NIHSS score was 3 points. The modified Rankin Scale (mRS) score was 1 point at 90-day telephone follow-up after thrombectomy. The patient has been taking dabigatran and atorvastatin regularly at home.

3. Discussion

Internal carotid artery congenital absence is a rare developmental abnormality. Some scholars believe that the congenital absence of ICA is a kind of neurocristopathy syndrome, which also includes neuroblastoma, pheochromocytoma, congenital hypoventilation syndrome, neurofibromatosis type I, DiGeorge syndrome, aortic coarctation, PHACE syndrome, and Moyamoyadisease (Bolande, 1974; Komiyama, 2020). From the perspective of embryology, the ICA begins to form when the embryo develops to the 3-mm stage. It comes from the first and third aortic arches and paired dorsal aorta. Some believe that ICA congenital absence is caused by disruption of the embryo by physical and haemodynamic pressure upon the embryo, and others consider it related to the abnormal degeneration of the first and third aortic arches.

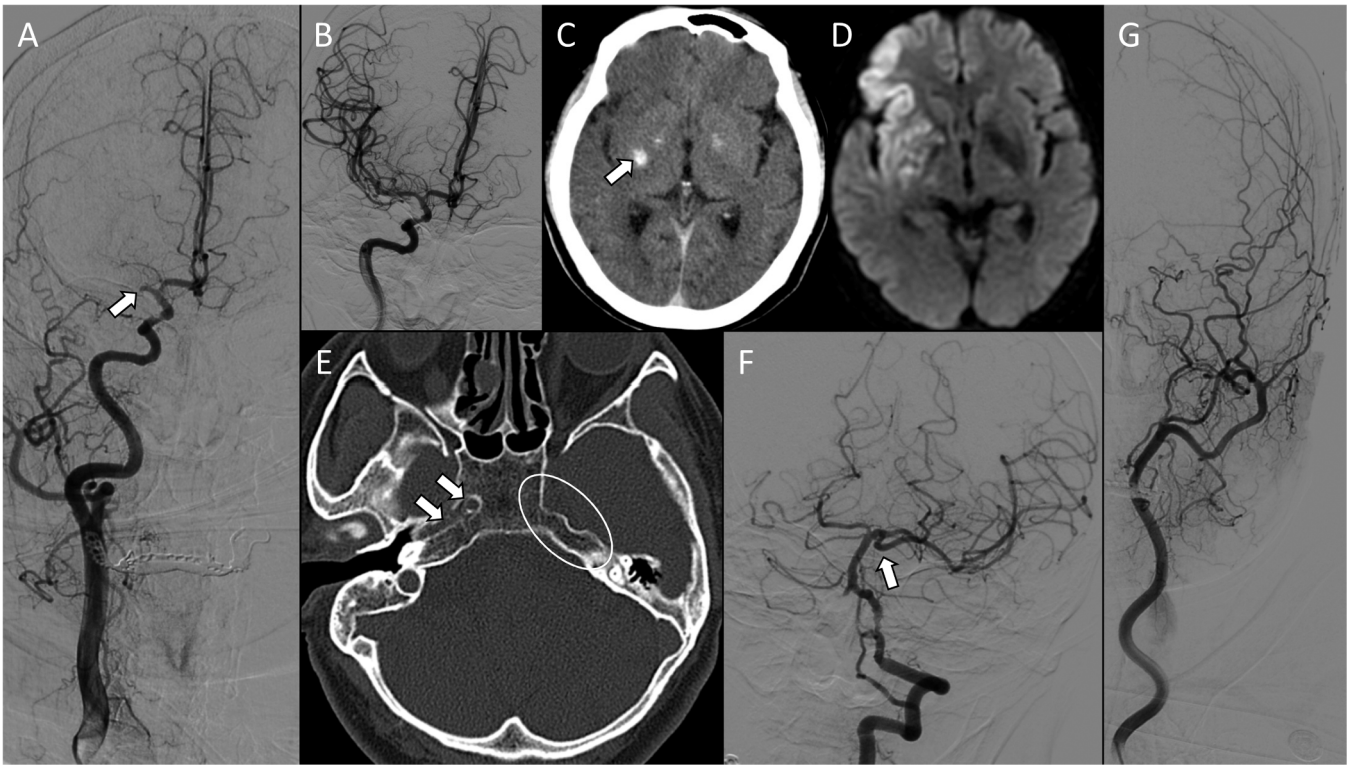

Fig. 1. Imaging of the perioperative period in this case. **A**:DSA indicated occlusion of the right MCA (white arrow), poor blood flow supplied by other arteries, and the left ACA on the side with the absent ICA was supplied by the ACoA; **B**: The patient had achieved complete reperfusion and direct aspiration via the distal access catheter, and the mTICI was grade 3; **C**: Immediate CT after thrombectomy showed a small high density in the right lenticular nucleus (white arrow), for which the CT value was 92 Hu considering exosmosis of contrast medium; **D**: MR-DWI at 48 h after thrombectomy showed that the diffusion of the frontal lobe, insular cortex and lenticular nucleus was limited unevenly; **E**: TheCT bone window showed that the morphology of the carotid canal in the petrous part of the right temporal bone was normal (white arrow), while the contralateral was absent (white oval); **F**: The blood-supplying region of the left middle cerebral artery was supplied by posterior circulation through the left thick PCoA (white arrow); **G**: The left CCA directly continued into the ECA, and no obvious collateral branches were formed to compensate for the blood supply to the brain parenchyma. Abbreviations:MCA,middle cerebral artery; ACA,anterior cerebral artery; ICA, internal carotid artery; ACoA,anterior communicating artery; PCoA,posterior communicating artery; CCA, commoninternal carotid artery; ECA, external carotid artery.

The carotid canal is a tubular structure within the petrous bone that is formed at 5–6 weeks of gestation and is dependent on the formation of the internal carotid artery. If the ICA does not develop, the ipsilateral carotid artery cannot form normally, though the exact cause is not clear (Kunishio et al., 1987; Cohen et al., 2010a). Referring to the standards of the 1996 Bouthillier seven segment classification (Bouthillier et al., 1996), the current diagnostic standard of congenital absence of the ICA is its complete absence and absence of the carotid artery. Dysplasia is the development of some segments of the ICA and the presence of the carotid canal. Therefore, the key way to distinguish whether the internal carotid artery is congenitally absent or has acquired occlusion is to confirm the presence or absence of carotid canal formation. That is, if the internal carotid artery is not formed during foetal life, the carotid canal will never form; if it is formed temporarily and then regresses early, the carotid canal is hypoplastic. Therefore, the existence of the carotid artery is the key point of differentiating ICA deficiency from congenital absence, dysplasia and chronic atherosclerotic occlusion. According to previous reports (Li et al., 2017), congenital absence of the ICA is mostly unilateral, and a few are bilateral, most of which (75%) are left congenital absence.

According to the images of the DSA and CT bone window of this case, the patient definitely hadcongenital absence of the left ICA. Lie and Hage (1968) divided ICA congenital absence into 6 types according to the different compensation methods of intracranial and extracranial blood vessels. In this case, the left anterior circulation mainly provided collateral blood supply through the ACoA and PCoA. The left anterior circulation cerebral blood supply was mainly provided by the right anterior circulation and left posterior circulation via the ACoA and PCoA. This collateral compensation method is in line with Type A of the Lie classification. The patients with ICA congenital absence always have no obvious clinical symptoms because of adequate collateral compensation. Although this patient had an in-hospital cardiogenic cerebral embolism due to atrial fibrillation and no anticoagulation therapy, the embolism was located in the right MCA trunk and avoided the A1 segment of the ipsilateral ACA. If the embolism had occurred in any segment of the right ICA, the compensatory mechanism via ACoA would have been disrupted, which would result in concurrent left frontal lobe ischaemic infarction and more severe neurological deficits. If the thrombus is located in the basilar artery or the P1 segment of the left posterior cerebral artery, the loss of compensatory mechanism via the PCoA can also lead to serious consequences, and interventional thrombectomy would be more difficult than that of the right anterior circulation.Due to sufficient compensation via the ACoA and PCoA, the left ECA fails to participate in the formation of pial collaterals via the artery or ophthalmic artery to supply blood to the brain parenchyma. Therefore, even if the left CCA or ECA is occluded, it is less likely to result in stroke.DSA indicated that the patient's right MCA trunk was acutely occluded, with poor collateral compensation in the ischaemic area. Fortunately, because the case was an in-hospital stroke, interventional thrombectomy bridging intravenous thrombolysis was given in a timely manner, and a good outcome was obtained.

Congenital absence of the ICA combined with large vessel disease in the contralateral anterior circulation is rarely reported worldwide. Due to the unique anatomical and haemodynamic differences in the congenital absence of an ICA, the morbidity of intracranial aneurysm in its complications can reach 27.8% (Zink et al., 2007), and there is also a combination of acute ischaemic stroke caused by atherosclerosis, transient ischaemic attack, pulsatile tinnitus, migraine, and Horner's syndrome caused by lesions (Cohen et al., 2010b; Hamada et al., 2021; Lee et al., 2012), but cases with cardiogenic cerebral embolism are very rarely reported.In this article, we provide a valuable typical case for reference by neurovascular interventional physicians. At the present day, the mechanical thrombectomy for intracranial artery occlusion caused by cardiogenic embolus is already being a routine interventional procedure. In this case, although the probability is relatively low, there is still a possibility that the cardiogenic embolus may enter the left vertebral artery and lead to the left middle cerebral artery (MCA) occlusion. Therefore, the value of this case is to show the anatomical variations and pathway changes of intracranial arteries which caused by the congenital ICA absence in a real stroke patient. If the embolism occurs at the proximal end of the left MCA in this patient, neurosurgeons or neuroradiologists should quickly identify and promptly change the endovascular treatment strategies.

4. Conclusion

We report a valuable case in which intravenous thrombolysis was ineffective against hyperacute stroke, with contralateral internal carotid artery congenital absence. In the cases of ICA congenital absence combined with hyperacute stroke in which intravenous thrombolysis is ineffective, interventional reperfusion operators should be familiar with the anatomical classification and the characteristics of the variants of the vasculature and should strive for an early diagnosis through the use of the CT bone window. The prognosis can be accurately judged according to the site of vascular occlusion so that appropriate intraoperative coping strategies can be adopted to improve the prognosis. In summary, we recognized that it is crucial to promptly and correctly identify the congenital vascular variations during the endovascular treatment through this case.

Funding

None.

Ethical statement

This article does not involve human or animal experiments, and all authors agree to publish it.

CRediT authorship contribution statement

G. Jin: Conceptualization, Formal analysis, Investigation, Methodology, Project administration, Resources, Writing – original draft, Writing – review & editing, Visualization. **J.W. Wang:** Investigation, Writing – original draft, Project administration. **Y. Zhang:** Investigation, Writing – original draft, Project administration. **X. Li:** Data curation, Formal analysis, Software. **Y.T. Yang:** Data curation, Formal analysis, Software. **Q.L. Zhan:** Conceptualization, Methodology, Project administration, Writing – review & editing.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

References

Bolande, R.P., 1974. The neurocristopathies: a unifying concept of disease arising in neural crest maldevelopment. Hum. Pathol. 5 (4), 409–429.
Bouthillier, A., Loveren, H.R.V., Keller, J.T., 1996. Segments of the internal carotid artery: anew classification. Neurosurgery 38 (3), 425–432.
Claros, P., Bandos, R., Gilea, I., et al., 1999. Major congenital anomalies of the internal carotid artery: agenesis, aplasia and hypoplasia. Int. J. Pediatr. Otorhinolaryngol. 49 (1), 69–76.
Cohen, J.E., Gomori, J.M., Leker, R.R., 2010a. Internal carotid artery agenesis: diagnosis, clinical spectrum, associated conditions and its importance in the era of stroke interventions. Neurol. Res. 32 (10), 1027–1032.
Cohen, J.E., Gomori, J.M., Leker, R.R., 2010b. Internal carotid artery agenesis: diagnosis, clinical spectrum, associated conditions and its importance in the era of stroke interventions. Neurol. Res. 32 (10), 1027–1032.
Hamada, Y., Ikeda, M., Yamashita, Y., et al., 2021. A 74-year-old man who underwent carotid artery stenting with symptomatic right internal carotid artery stenosis with congenital agenesis of the left internal carotid artery. RinshoShinkeigaku 61 (10), 696–699.

Komiyama, M., 2020. Segmental vulnerability and vascular neurocristopathy of the internal carotid artery. Interv. Neuroradiol. 26 (2), 131–134.
Kunishio, K., Yamamoto, Y., Sunami, N., et al., 1987. Agenesis of the left internal carotid artery, common carotid artery, and main trunk of the external carotid artery associated with multiple cerebral aneurysms. Surg. Neurol. 27 (2), 177–181.
Lee, E.J., Chang, H.W., Ha, E.Y., et al., 2012. Acute infarction in a patient with agenesis of the internal carotid artery and transcavernous anastomosis. J. Clin. Neurosci. 19 (4), 597–599.
Lee, J.H., Oh, C.W., Lee, S.H., et al., 2003. Aplasia of the internal carotid artery. Acta Neurochir. 145 (2), 117–125.
Li, S., Hooda, K., Gupta, N., et al., 2017. Internal carotid artery agenesis: a case report and review of literature. Neuroradiol. J. 30 (2), 186–191.
Lie, T.A., Hage, J., 1968. Congenital anomalies of the carotid arteries. Plast. Reconstr. Surg. 42 (3), 283.
Midkiff, R.B., Boykin, M.W., McFarland, D.R., et al., 1995. Agenesis of the internal carotid artery with intercavernous anastomosis. Am. J. Neuroradiol. 16 (6), 1356–1359.
Zink, W.E., Komotar, R.J., Meyers, P.M., 2007. Internal carotid aplasia/hypoplasia and intracranial saccular aneurysms: series of three new cases and systematic review of the literature. J. Neuroimaging 17 (2), 141–147.