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Case Report

Unilateral agenesis of the internal carotid artery with intercavernous anastomosis combined with an unruptured internal carotid artery aneurysm

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Abstract: Unilateral agenesis of the internal carotid artery (ICA) with intercavernous anastomosis is extremely rare. In addition, of these rare cases, only 4 patients had combined intracranial aneurysms, which were located in the anterior communicating artery and posterior cerebral artery. The onset symptoms of these 4 patients included headache, ischemic stroke and aneurysm rupture, but none of them showed intracerebral hemorrhage at onset. This is the first case of unilateral agenesis of the ICA with intercavernous anastomosis combined with an unruptured aneurysm located in the ICA, which had a spontaneous thalamic hemorrhage as the onset symptom. Based on the digital subtraction angiography (DSA) images of the patient, we speculated that asymmetric distribution of blood flow in the posterior circulation secondary to agenesis of the ICA might be a cause of thalamic hemorrhage. Given the presence of an intact aneurysm and the influence of agenesis of the ICA on cerebral blood perfusion, strict follow-up is critical for these patients.

Keywords: Agenesis of internal carotid artery, intercavernous anastomosis, aneurysm, thalamic hemorrhage, ischemic stroke

Introduction

Unilateral agenesis of the internal carotid artery (ICA) is a rare vascular abnormality with an incidence of less than 0.01% [1]. The disease is typically identified during head and neck imaging examinations, but in some cases, the clinical symptoms could be caused by cerebrovascular insufficiency, compression by enlarged intracranial collateral vessels (or aneurysm), or spontaneous subarachnoid hemorrhage. Based on different compensatory pathways of collateral circulation, Lie et al. classified ICA agenesis into six types, type A-F [2]. Among them, ICA agenesis with an abnormal intercavernous anastomosis (type D) is particularly rare (26 cases reported), and only 4 cases combined with intracranial aneurysms have been reported to date [3-6]. Here, we report a case of unilateral agenesis of the ICA with a collateral supply of blood through an anastomotic vessel arising from the cavernous region of the contralateral ICA, which is combined with an unruptured ICA aneurysm. Among all 5 cases (including the one reported herein), this is the first case with an aneurysm arising from the ICA and the first case with spontaneous thalamic hemorrhage as the onset symptom.

Case report

A 64-year-old female visited our hospital in March 2018 due to sudden dizziness accompanied by numbness of the left side of her face and left limbs for two hours. The patient had no history of hypertension, diabetes, long-term medication, or head or neck trauma. A neurological examination suggested a sensory disturbance of the left limbs and normal muscular strength. A laboratory examination showed normal coagulation function.

A cranial CT suggested a hemorrhage of 2.2 × 1.8 × 1.2 cm in the right thalamus. The skull base CT scan revealed a complete absence of the right carotid canal (**Figure 1**). Digital sub-

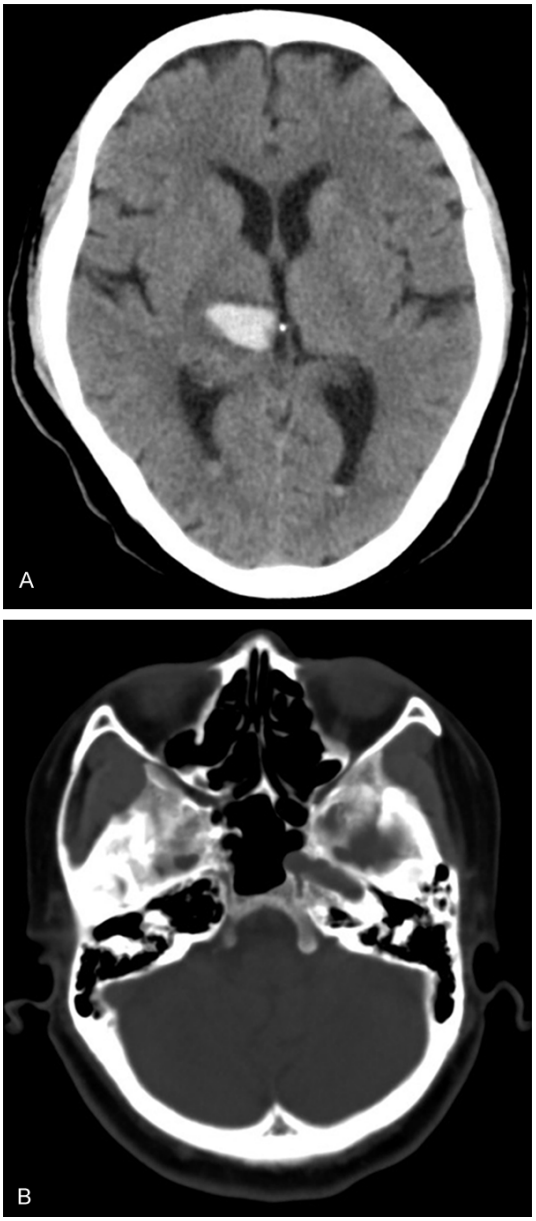

Figure 1. Cranial CTA. A. Slight hemorrhage in the right thalamus; B. Skull base CT suggested the absence of the right carotid canal.

traction angiography (DSA) did not identify the proximal end of the cavernous sinus of the right ICA. The cavernous sinus of the right ICA and its distal end were supplied by the anastomotic artery derived from the left ICA, and this artery crossed the midline and connected the cavernous sinuses of the bilateral ICA. The A1 segment of the right anterior cerebral artery (ACA) was absent. 3D-DSA revealed a 0.6 × 0.6 cm, wide-necked aneurysm in the lacerum segment (C3) of the left ICA. The right external carotid artery (ECA) was well-developed, but the right ICA was absent, and no right carotid bifurcation was identified (**Figure 2**).

A definite diagnosis was made as follows: 1) spontaneous right thalamic hemorrhage; 2) agenesis of the right ICA with intercavernous anastomosis; 3) aneurysm in the lacerum segment of the left ICA.

The patient was given conservative therapy during seven days of hospitalization, and then discharged after her symptoms improved. The patient was followed up for six months with no adverse events.

Discussion

The rarity of ICA agenesis with intercavernous anastomosis combined with an intracranial aneurysm

Unilateral agenesis of the ICA is a rare vascular abnormality with an incidence of less than 0.01%. The low incidence could be because the occurrence of such a vascular variation is inherently very low, and most patients with unilateral agenesis of the ICA would not develop clinical symptoms due to sufficient compensation of cerebral blood flow [7]. Embryologically, the proximal ICA develops bilaterally from the third dorsal aortic arch. It is postulated that agenesis of the ICA is due to mechanical and hemodynamic stresses placed on the embryo, such as exaggerated embryo folding in one plane and amniotic band constriction [8]. Since the carotid canal develops secondary to the fetal ICA, the CT finding of no carotid canal confirms true agenesis rather than aplasia or hypoplasia [9]. Some previous cases with an absence of the ICA had a cross-midline anastomosis between the bilateral ICA (type D) but lacked the CT finding of no carotid canal. Hence, they are not included in this review as cases of agenesis of the ICA. Among the reported 26 cases of type D agenesis of the ICA, there were only 4 cases combined with an intracranial aneurysm (**Table 1**). Three combined aneurysms arising from the anterior communicating artery, with the other one arising from the posterior cerebral artery. So, our case would be the first instance of a type D agenesis of the ICA combined with an aneurysm arising from the ICA (the lacerum segment of the ICA, an aneurysm at this site is extremely rare among the general population). The percentage of cases combined with aneurysms among all 27 cases of type D ICA agen-

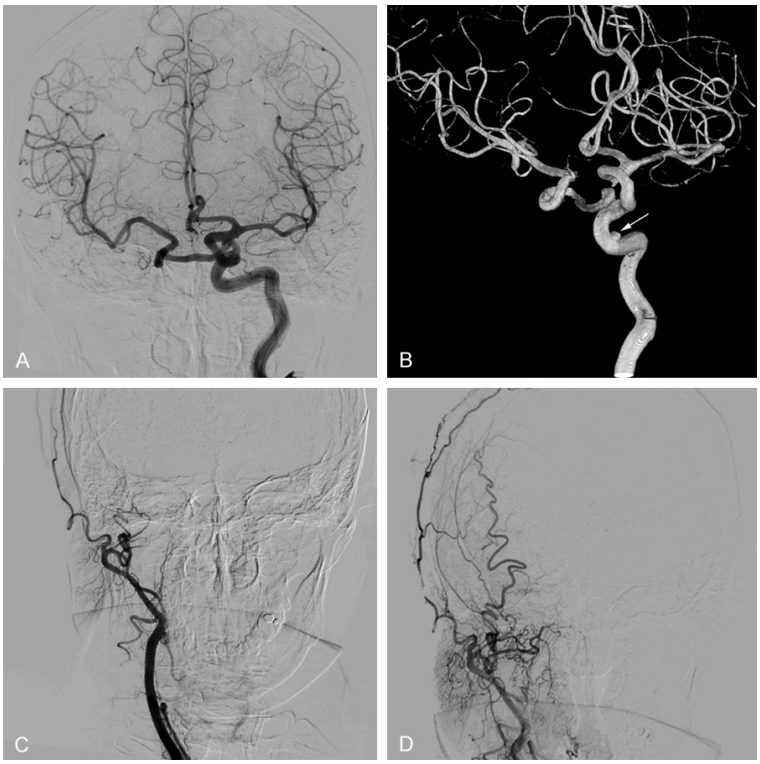

Figure 2. DSA imaging. A. DSA showed the absence of the proximal end of the cavernous sinus of the right ICA, the absence of the A1 segment of the right anterior cerebral artery, and visible anastomotic arteries crossing the midline and connecting the cavernous sinuses of the bilateral ICA; B. 3D-DSA showed an aneurysm in the lacerum segment of the left ICA (white arrow); C. Right carotid angiography shows no development in the right carotid bifurcation; D. The anastomotic vessels between extracranial and intracranial arteries for blood compensation were also absent.

esis (including our patient) was 18.5% (5/27), which is also much higher than the incidence of cerebral aneurysms among the general population (2-4%). This might be associated with abnormal embryonic development or caused by hemodynamic derangement [10].

The special onset symptom of our case

To the best of our knowledge, this is the first case report of a type D agenesis of the ICA with spontaneous thalamic hemorrhage as the onset symptom. The patient neither had a history of hypertension or diabetes, nor did she take anticoagulant or antiplatelet drugs. She showed normal coagulation function. Therefore, the hemorrhage was considered to be closely related to the change of cerebral arterial blood supply and hemodynamic derangement. The patient's DSA image of the vertebral-basilar artery indicated that when compared with the left side, the feeding area of the posterior cerebral artery on the right side had more enriched vessels and intensive staining (**Figure 3**). Thus, we speculate that such asymmetric distribution of blood flow, which was a compensation for agenesis of the right ICA could be the cause of spontaneous thalamic hemorrhage.

The clinical significance of agenesis of the ICA

Patients with agenesis of the ICA may suffer from stroke due to insufficient cerebral blood perfusion. Therefore, for these patients, lifestyle modifications and control of the risk factors for stroke are particularly important. Additionally, for those considering ICA ligation, carotid endarterectomy or transsphenoidal surgery, agenesis of the ICA must be excluded before operation due to potential disastrous consequences. Lastly, patients with agenesis of the ICA require strict follow-up. The follow-up would help to observe an existing or new aneurysm, and the patients may show ischemic symptoms with the progression of atherosclerosis [11], which might require an EC-IC bypass [1].

Conclusion

We presented the first case of unilateral agenesis of the ICA with intercavernous anastomosis combined with an unruptured ICA aneurysm. This is also the first case of agenesis of the ICA with spontaneous thalamus hemorrhage as an onset symptom. We suggest that patients with agenesis of the ICA be strictly followed up due to the risk of stroke and to detect any existing intracranial aneurysms.

Acknowledgements

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Table 1. Cases of unilateral agenesis of the internal carotid artery with intercavernous anastomosis combined with intracranial aneurysms

No.	Author	Year	Age	Gender	Side of ICA agenesis	Absence of Ipsilateral A1	Aneurysm	Symptoms at admission	plain CT findings	Other information
1	Quint DJ [3]	1989	65	Female	Right	Yes	Acom	Sudden onset severe headache	SAH	
2	Bodhey NK [4]	2004	57	Male	Right	Yes	PCA	None	None	Aberrant origin of the right subclavian artery
3	Horie N [5]	2008	55	Female	Left	Yes	Acom	None	None	Acom fenestration
4	Kumagai K [6]	2017	47	Male	Left	Yes	Acom	Numbness of left hand	None	
5	Present case	2018	64	Female	Left	Yes	ICA Lacerum segment	Left limb numbness	Thalamic hemorrhage	

Note: NA, not available; Acom, anterior communication artery; PCA, posterior cerebral artery. ICA, internal carotid artery; SAH, subarachnoid hemorrhage.

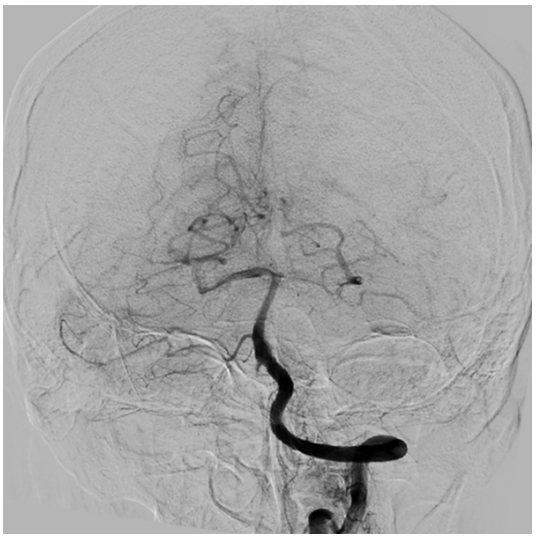

Figure 3. DSA of the vertebral-basilar artery. Compared to the left side, the feeding area of the posterior cerebral artery on the right side had more enriched vessels and intense staining.

Disclosure of conflict of interest

None.

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