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Bilateral Internal Carotid Artery Agenesis Except Unilateral Terminal Internal Carotid Artery Remnant

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Abstract

Bilateral internal carotid artery (ICA) agenesis is an extremely rare congenital anomaly. We present an unusual case of bilateral ICA agenesis except for a unilateral terminal ICA remnant. A 53-year-old woman presented with sudden severe headache. Brain computed tomography demonstrated left hemisphere cortical hemorrhage and subarachnoid hemorrhage. Cerebral angiography showed absence of bilateral ICA except for a left ICA terminal segment. Collateral circulation to anterior circulation was supplied from right carotid rete mirabile from the ascending pharyngeal artery, and a basilar artery supplying anterior circulation via enlarged POCMAs. The right vertebral artery was connected with the ascending cervical artery from the right subclavian artery and the right occipital artery. To our knowledge, this anomaly has not yet been reported in the literature. We consider that the increased turbulence and shearing stress in the intracranial circulation may have resulted in subarachnoid hemorrhage.

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Agenesis, Carotid rete mirabile, Collateral circulation, Internal carotid artery

Introduction

Agenesis, aplasia, and hypoplasia of the internal carotid artery (ICA) are rare congenital anomalies, which occur in less than 0.01% of the population [4,12]. Unilateral ICA agenesis is more common than bilateral ICA agenesis [3,7]. In cases of bilateral ICA agenesis, anterior circulation is usually supplied by an enlarged posterior communicating artery (POCMA) from a basilar artery (BA) or a carotid rete mirabile from the external carotid artery [7]. To our knowledge, bilateral ICA agenesis except for a unilateral terminal ICA with collateral flow via both POCMAs and a unilateral carotid rete mirabile has not yet been reported in the literature. Here we present an unusual case of bilateral ICA agenesis except for a unilateral terminal ICA remnant, which is associated with a unilateral carotid rete mirabile from the ascending pharyngeal artery and unilateral VA augmented by an occipital artery and an ascending cervical artery.

Case Report

A 53-year-old woman presented with sudden severe headache three hours previously. Neurological examination at another hospital showed headache, slightly drowsy mentality, and neck stiffness.

Past medical history: Twenty-seven years earlier, the patient underwent correction for tetralogy of Fallot at other University hospital. Eight years, earlier she underwent percutaneous coronary intervention for her right coronary artery because of angina pectoris in our hospital. She had taken escitalopram oxalate (Lexapro, Lundbeck Korea Co, Seoul, South Korea) 5 mg, Astrix (aspirin, BoRyung Pharmaceutical, Seoul, South Korea) 100 mg, furosemide (Lasix, Handok, Seoul, South Korea) 20 mg, spironolactone (Aldactone, film coated, Pfizer Pharmaceutical Korea, Seoul, South Korea) 12.5 mg po once a day. She had taken trimetazidine dihydrochloride (Vastinan SR, Servier Korea, Seoul, South Korea) 35mg, cilostazol (Pletaal, Korea Otsuka Pharmaceuticals, Seoul, South Korea) 50 mg po twice a day. Brain magnetic resonance (MR) imaging performed at outside hospital six years earlier demonstrated no signal void in the right cavernous sinus and a suspicious signal void in the left cavernous sinus. Brain computed tomographic angiography (Aquilion Prime 160-slice CT scanner, Toshiba Medical Systems, Tokyo, Japan) performed after iopamidol (Pamiray 370, Dongkook Pharmaceuticals, Seoul, Korea) injection three years earlier demonstrated tortuous BA and enlarged both POCMAs, especially the long right one (Figure 1). MR angiography of the neck (Signa HDx, GE, USA) performed after ProHance (Gadoteridol, Bracco Diagnostic Inc, Monroe Township, NJ) injection two years earlier showed that the right VA was the main supply of intracranial circulation (Figure 2).

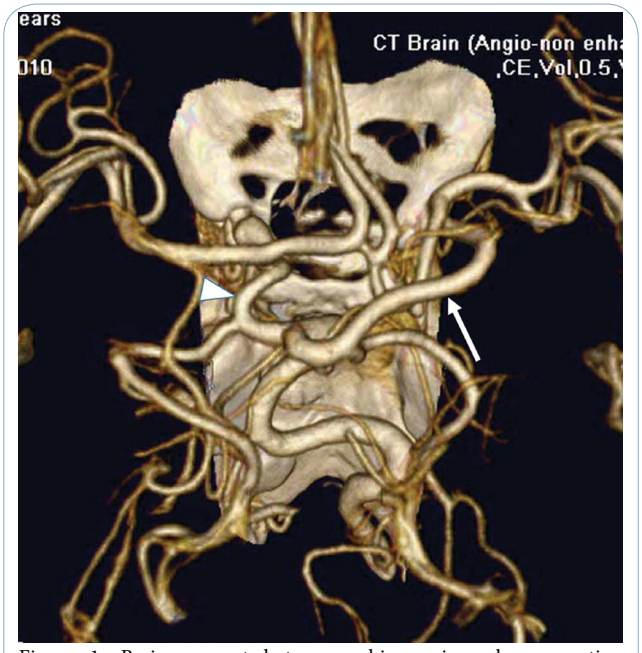

Figure 1: Brain computed tomographic angiography presenting a tortuous basilar artery and enlarged posterior communicating arteries bilaterally, especially long on the right. (Arrow: right posterior communicating artery, arrowhead: left posterior communicating artery).

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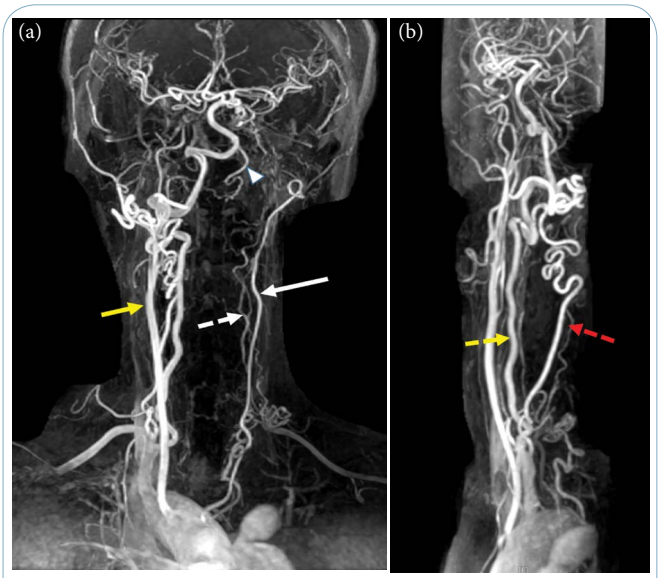

Figure 2: Magnetic resonance angiography of the neck (a, b) demonstrating that the right vertebral artery was the main supply of intracranial circulation. The left common carotid artery (white arrow) originates from the aortic arch but does not have a left internal carotid artery. The left vertebral artery (white dotted arrow) from the left subclavian artery does not have a definite intracranial supply. The left V4 segment (white arrowhead) has collateral flow from the right vertebral artery and the right common carotid artery. The right common carotid artery (yellow arrow) originates from the aortic arch and has a suspicious intracranial supply. The right vertebral artery (yellow dotted arrow) is connected with the ascending cervical artery (red dotted arrow) from the right subclavian artery and supplies the intracranial circulation.

Imaging study: Brain computed tomography (CT) in the emergency department at outside University hospital demonstrated left hemisphere cortical hemorrhage and subarachnoid hemorrhage (Figure 3). Cerebral angiography the following day at outside University hospital showed absence of bilateral ICA except for a left ICA terminal segment, a right carotid rete mirabile from the ascending pharyngeal artery, and a BA supplying anterior circulation via enlarged POCMAs. The right vertebral artery (VA) was connected with the ascending cervical artery from the right subclavian artery and the right occipital artery (Figures 4-7). Bone setting brain CT demonstrated the absence of both carotid canals (Figure 8). The patient was treated conservatively and was discharged with memory disturbance and a language problem. All studies and procedures in our hospital were performed after acceptance of informed consent.

Follow-up: Three years subsequently, the patient still showed memory disturbance, anxiety, and a language problem (mild aphasia with difficulty in word finding). However, these deficits showed no change during three years.

Discussion

Type of ICA agenesis

Lie [11] described six pathways of collateral circulation in association with the absence of the ICA. In type A, at the side of the malformation, an enlarged POCMA supplies the ipsilateral middle cerebral artery (MCA), and the anterior cerebral artery (ACA) is supplied through the anterior communicating artery. Type B refers to a patent anterior communicating artery that supplies the ACA and MCA. Type C occurs when both internal carotids are absent and the

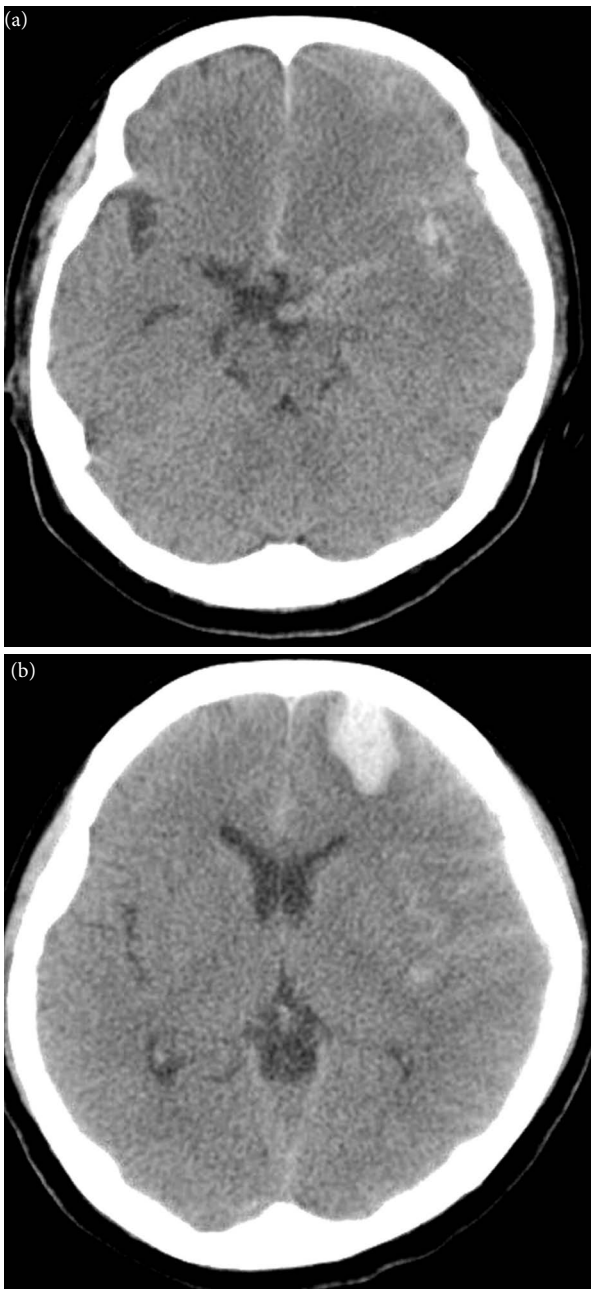

Figure 3: Brain computed tomography (a, b) in the emergency department demonstrating a left hemisphere cortical hemorrhage and subarachnoid hemorrhage located mainly in the left hemisphere.

ACA and MCA are supplied by vessels from the BA. In type D, the cervical and petrous portions of the unilateral ICA are absent, and blood supply is from the intercavernous communication. In type E, bilateral ACAs are supplied by hypoplastic ICAs, and bilateral MCAs are supplied by enlarged POCMAs. In type F, the precavernous portion of the ICA is formed by several arteries constituting a rete mirabile that fills the cavernous portion in the absence of the proximal portions of the ICA.

Types A, B, and D are unilateral agenesis of the ICA. Type C is bilateral ICA agenesis. Type E is hypoplasia of the ICA. Type F is unilateral or bilateral agenesis of the ICA. The present patient had a BA suppling anterior circulation via the POCMA. Therefore, her case

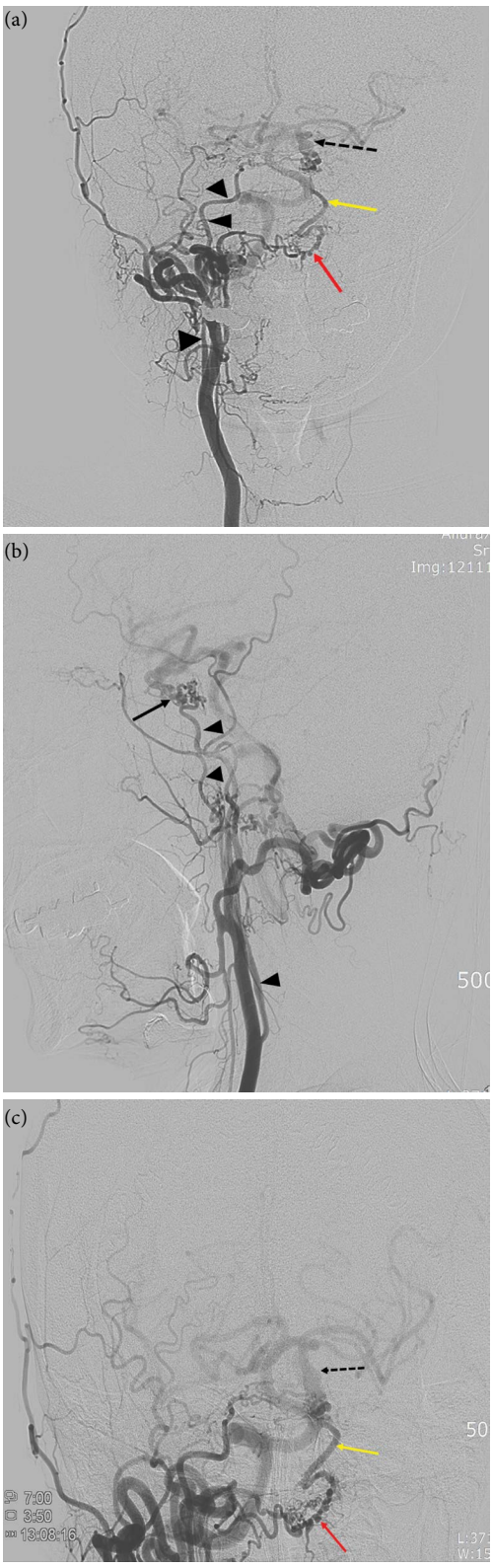

Figure 4: Right common carotid arteriogram (a, b, c). The right common carotid arteriogram showing no right internal carotid artery. This angiogram demonstrates a right carotid rete mirabile (black arrow) from the ascending pharyngeal artery (black arrowhead) and a dilated left ICA terminal segment (black dotted arrow). The right external carotid artery has collateral flow (red arrow) to the left vertebral artery V4 segment (yellow arrow).

is similar to Type C. However, this case has several characteristics distinct from type C. First, the patient had a right carotid rete mirabile originating from the ascending pharyngeal artery, and she had a right VA augmented by the right ascending cervical artery and right occipital artery. Moreover, the patient had a cardiac anomaly. All reported patients with cases of type C ICA agenesis had VAs that were both normal [1-2, 8-10, 13-21]. Our patient had a unilateral normal VA and a contralateral left terminal VA. To our knowledge, this anomaly has not yet been reported in the literature.

The diagnosis of congenital absence of the ICA can be made by skull base CT. The finding on a routine CT of an absent or hypoplastic carotid canal is a radiological method of congenitally anomalous ICA diagnosis [6]. In this case, we identified absence of both carotid canals.

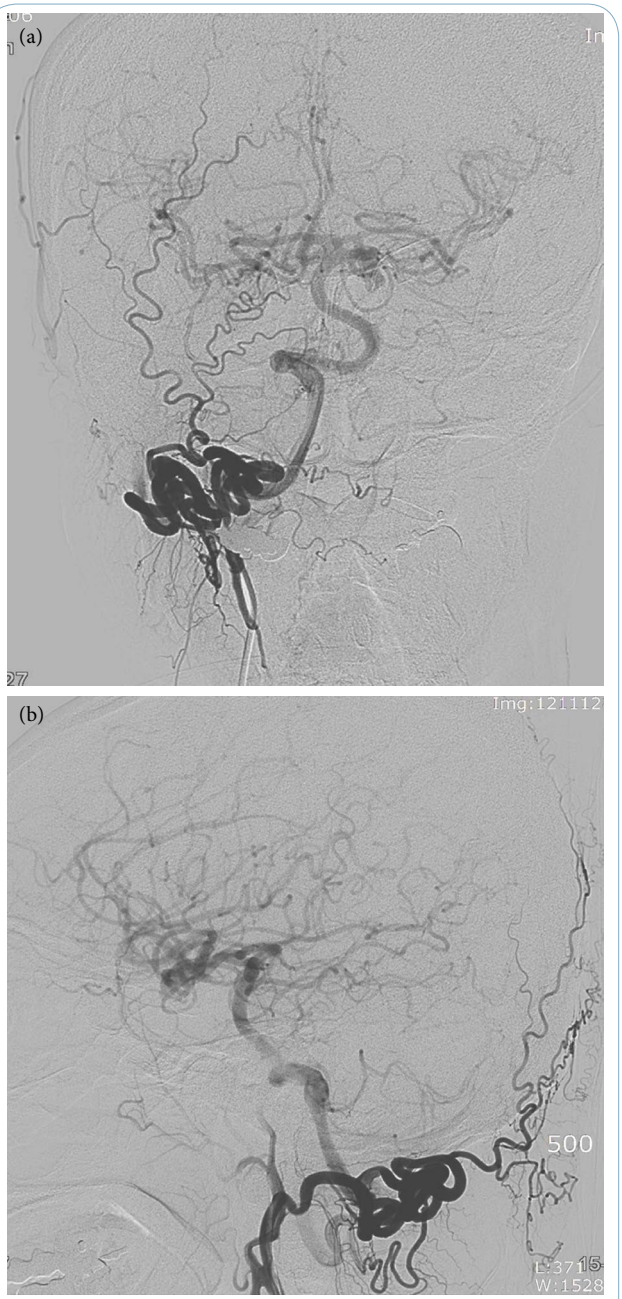

Figure 5: Right external carotid arteriogram (a, b) showing that the right vertebral artery has collateral flow via the right occipital artery.

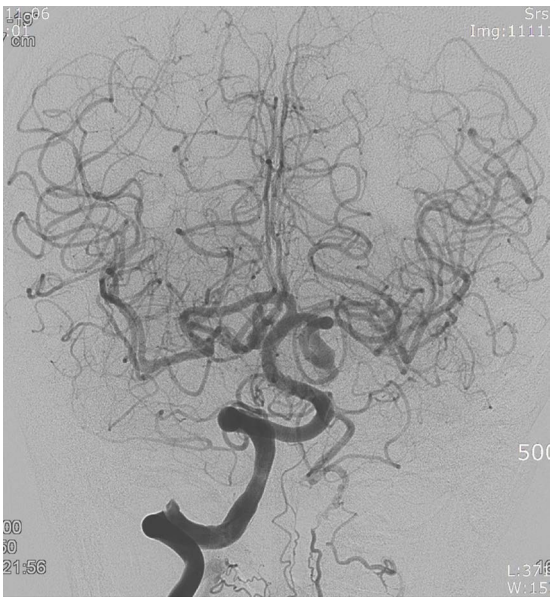

Figure 6: Right distal vertebral angiography demonstrating a tortuous basilar artery supplying the anterior circulation via a posterior communicating artery.

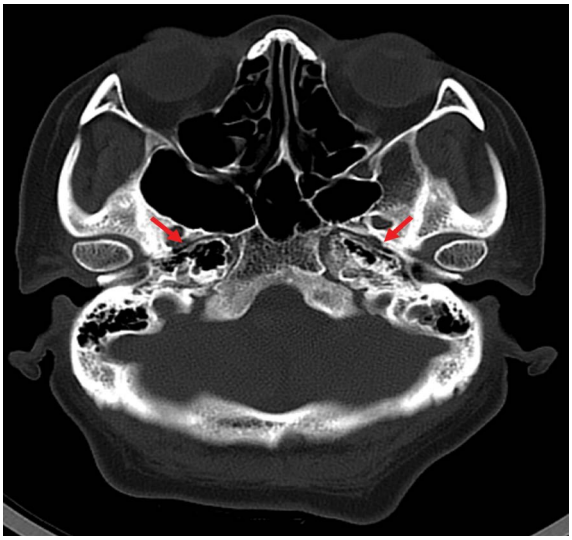

Figure 8: Bone setting computed tomography demonstrating absence of bilateral carotid canals (red arrow).

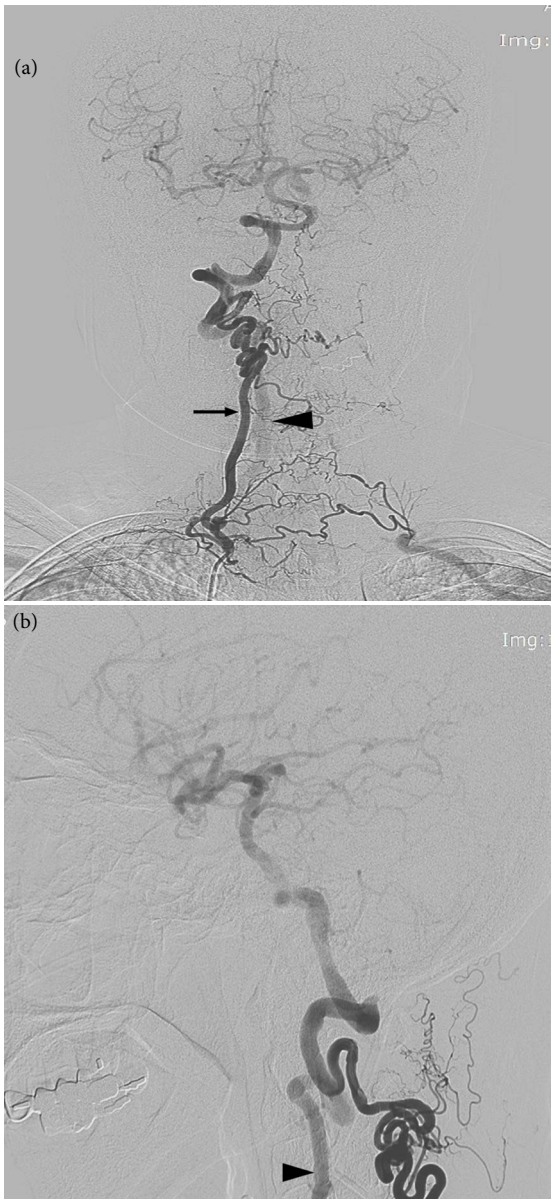

Figure 7: Contrast injection into the ascending cervical artery (arrow) showing connection with the original right vertebral artery (arrowhead).

Symptom of Patients with Bilateral ICA Agenesis

Most patients with a congenital absence of ICA are detected incidentally by cerebral angiography performed for other reasons. Usually they have no neurological deficit because of a good collateral blood supply to the affected side [22]. When present, the symptoms may be the result of cerebrovascular insufficiency in a cerebral area, compression of brain parenchyma by enlarged vessels, or an aneurysm [5, 16]. Zink et al. [22] reported in their review of 199 cases of unilateral and bilateral ICA aplasia/hypoplasia that signs and symptoms attributable to acute ischemia were most common, including transient ischemic attack, stroke, amaurosis fugax, or ischemic optic neuropathy.

Review of type C ICA agenesis cases demonstrated that presenting symptoms were cerebellar hemorrhage [9,17], subarachnoid hemorrhage from aneurysm [2], mental retardation and developmental delay [10,13,21], chest pain and fainting spells, headache [1,14,16,18,20], and transient ischemic attack [3,19]. In this case, the right VA is the main blood supply for the entire brain circulation. However, this blood supply may not be sufficient for brain demand. Therefore, both the carotid rete mirabile from the right ascending pharyngeal artery and the right VA augmented by the right occipital artery and right ascending cervical artery supplemented intracranial blood supply. However, in this case, the increased turbulence and shearing stress may have resulted in subarachnoid hemorrhage and cortical hemorrhage not originating from aneurysm. This subarachnoid and cortical hemorrhage in type C ICA agenesis maybe unreported presenting symptom.

Recognition of bilateral ICA agenesis is important for several reasons. First, cerebral thromboembolic disease of anterior circulation

can be explained by atherosclerotic disease of the vertebrobasilar system. Second, a poor collateral supply or abnormal location of vessels to the head places patients at high risk during catheter angiography and surgery. Finally, there is a strong association for the development of cerebral aneurysm because of hemodynamic stress [19]. Additionally, subarachnoid hemorrhage may result not from an aneurysm but from hemodynamic compromise, as in this case.

Conclusion

We reported bilateral ICA agenesis except for a unilateral terminal ICA remnant. In the present case, collateral flow to the anterior circulation was supplied from both a unilateral carotid rete mirabile and a BA via enlarged POCMAs. The patient had a cardiac anomaly and an abnormally developed left VA. We consider that the increased turbulence and shearing stress in the intracranial circulation may have resulted in subarachnoid hemorrhage.

Competing Interests

The authors have no competing interests with the work presented in this manuscript.

Acknowledgments

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