Acute Cerebral Infarction in a Patient with Persistent Trigeminal Artery and Homolateral Hypoplasia of Internal Carotid Artery Distal Anastomosis: A Case Report and a Mini Review of the Literature

Da-Ping Lyu, MD,* Yu Wang, MD, PhD,† Kai Wang, MD, PhD,† Ming Yao, MD,* Yun-Fan Wu, PhD,‡ and Zhi-Hua Zhou, PhD,§

Background: Persistent primitive trigeminal artery (PPTA) is a rare remnant between the internal carotid artery (ICA) and the basilar artery into adulthood. PPTA generally lacks specific clinical manifestations and occasionally accompanies with other cerebrovascular diseases. *Case presentation:* We reported a 48-year-old Chinese woman who had repeated episodes of transient ischemic attack presented to our hospital. She had no related risk factors of ischemic cerebrovascular diseases. Magnetic resonance image findings demonstrated acute cerebral infraction in centrum semiovale. Magnetic resonance angiography findings indicated right PPTA and ipsilateral hypoplasia of ICA distal anastomosis. *Conclusions:* To the best of our knowledge, this is the first report that acute cerebral infarction in a patient with the right PPTA and ipsilateral hypoplasia of ICA distal anastomosis. According to the literature, congenital factor may play an important role in the formation of these vascular anomalies.

Key Words: Persistent primitive trigeminal artery (PPTA)—internal carotid artery (ICA)—basilar artery (BA)—magnetic resonance angiography (MRA)—acute cerebral infarction

Introduction

Multiple fetal arterial anastomoses exist between the internal carotid artery (ICA) and the vertebrobasilar artery system during the early stages of embryogenesis. Four pairs of fetal anastomosis were described which include the trigeminal, otic, hypoglossal, and proatlantal intersegmental arteries between the anterior and posterior circulation.[1,2] All these primitive arterial connection regresses and then disappear when definitive cerebral circulation forms. Persistent primitive trigeminal artery (PPTA) is a rare remnant in the embryonic circulatory system which arches the proximal cavernous segment of the ICA with the middle or distal portion of the basilar artery (BA) into adulthood. PPTA accounts for the most common anastomosis between the ICA and vertebrobasilar artery. PPTA generally lacks specific clinical presentations and occasionally accompanies with other cerebrovascular anomalies including aneurysm,[3] cerebrovascular malformation,[4] moyamoya disease,[5] transient ischemic attack .[6]

As far as we know, acute cerebral infarction in a patient with right PPTA and the ipsilateral hypoplasia of ICA distal

Abbreviations: PPTA, persistent primitive trigeminal artery; ICA, internal carotid artery; BA, basilar artery; VA, vertebral artery; MRI, magnetic resonance imaging

From the *Department of Neurology, The Fourth Affiliated Hospital of Anhui Medical University, Hefei, Anhui, China; †Department of Neurology, The First Affiliated Hospital of Anhui Medical University, Hefei, Anhui, China; ‡Department of Medical Imaging, Guangdong Second Provincial General Hospital, Guangzhou, Guangdong, China; and §Department of Neurology, The First Affiliated Hospital, School of Clinical Medicine of Guangdong Pharmaceutical University, Guangzhou, Guangdong, China.

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Address correspondence to Zhi-Hua Zhou, PhD, Department of Neurology, The First Affiliated Hospital, School of Clinical Medicine of Guangdong Pharmaceutical University, Guangzhou, Guangdong, China and Address correspondence to Yu Wang, MD, PhD, Department of Neurology, The First Affiliated Hospital of Anhui Medical University, Hefei, Anhui, China. E-mails: wangyu18b@163.com, zhouzh20051103@163.com.

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anastomosis are not reported. Here we present a female patient who was assessed for acute centrum semiovale infarction and cranial magnetic resonance angiography (MRA) findings demonstrate a right PPTA with hypoplasia of the ipsilateral ICA distal anastomosis. Due to the MRA findings in this case, we discuss and briefly review it.

Case Presentation

A 48-year-old Chinese woman was referred to our hospital with weakness in her left upper extremity and dizziness for the past 18 hours. Over the past 18 hours, these symptoms had been occurred 3 times and each time lasting 2 minutes. The patient had no medical history of hypertension, diabetes mellitus, dyslipidemia, heart disease, or blood disease. On admission, she was conscious but with normal verbal fluency. Her vital signs are stable (body temperature was 36.2°C, blood pressure was 110/70 mm Hg, pulse rate was 70 beats/minute). Cardiovascular, respiratory, and abdominal examinations were unremarkable, while thorough neurologic assessment revealed a positive Barré test on his right upper extremity. Laboratory findings, such as fasting blood glucose, HbA1C, blood lipids, uric acid, homocysteine, coagulation parameters, protein C/S, NT-pro-BNP, autoantibody, were well within the normal reference range. Detailed results were shown in Table 1. Twenty four hour Holter electrocardiography showed sinus rhythm without ST-T segment change and occasional premature atrialcontraction (15 times within 24 hours). Echocardiography showed

Table 1. *Laboratory findings in the patient*

Laboratory tests	Results	Normal range
Fasting blood sugar	4.8 mmol/L	3.89-6.1 mmol/L
2h postprandial blood sugar	6.8 mmol/L	<7.8 mmol/L
HbA1c	5.6%	4-6%
Total cholesterol (TC)	4.35 mmol/L	3.5-5.69 mmol/L
Triglyceride (TG)	1.66 mmol/L	.45-1.70 mmol/L
Low-density lipoprotein cholesterol (LDL-c)	3.10 mmol/L	<3.12 mmol/L
High-density lipoprotein cholesterol (HDL-c)	.72 mmol/L	.7-2.0 mmol/L
Platelets count	128×10^9/L	$100\text{-}300 \times 10^9$/L
NT-pro-BNP*	109.4 pg/ml	0-300 pg/ml
Free triiodothyronine (FT3)	3.38 pmol/L	2.63-5.7 pmol/L
Free tetraiodothyronine (FT4)	12.20 pmol/L	9.01-19.05 pmol/L
Thyroid stimulating hormone (TSH)	2.26 μIU/ml	.35-4.94 μIU/L
antithyroglobulin antibody (TGAb)	.69 IU/mL	0-4.11 IU/mL
Thyroid peroxidases antibody (TPOAb)	.72 IU/mL	0-5.61 IU/mL
Prothrombin time (PT)	13.2 s	11.0-15.0 s
Activated partial thromboplastin time (APTT)	31.4 s	28.0-45.0 s
Thrombin time (TT)	15.7 s	14.0-21.0 s
International normalized ratio (INR)	1.11	.8-1.2
Protein C	94%	60-140%
Protein S	97%	63.5-149%
Complement 3 (C3)	1.35 g/L	.85-1.93 g/L
Complement 4 (C4)	.26 g/L	.12-.36 g/L
Anti-streptolysin O (ASO)	54.1 IU/mL	.1-200 IU/mL
Rheumatoid factor (RF)	12.5 IU/L	.1-15 kU/L
Hypersensitivity C reactive protein (hs-CRP)	2.57 mg/L	0-3.0 mg/L
Antinuclear antibody (ANA)	.3 u/ml	0-18 u/ml
Anti-SS-A antibody	Negative	Negative
Anti-SS-B antibody	Negative	Negative
Anti-Sm antibody	Negative	Negative
Anti-Ro-52 antibody	Negative	Negative
Anti-dsDNA antibody	Negative	Negative
Anti-dsDNA antibody	Negative	Negative
Anti-centromere antibody	Negative	Negative
Anti-nucleosome antibody	Negative	Negative
Type 2 anti-mitochondrial antibody	Negative	Negative
Anti-cardiolipin antibody IgA	Negative	Negative
Anti-cardiolipin antibody IgG	Negative	Negative
Anti-cardiolipin antibody IgM	Negative	Negative

*NT-pro-BNP: N-terminal fragment brain natriuretic peptides.

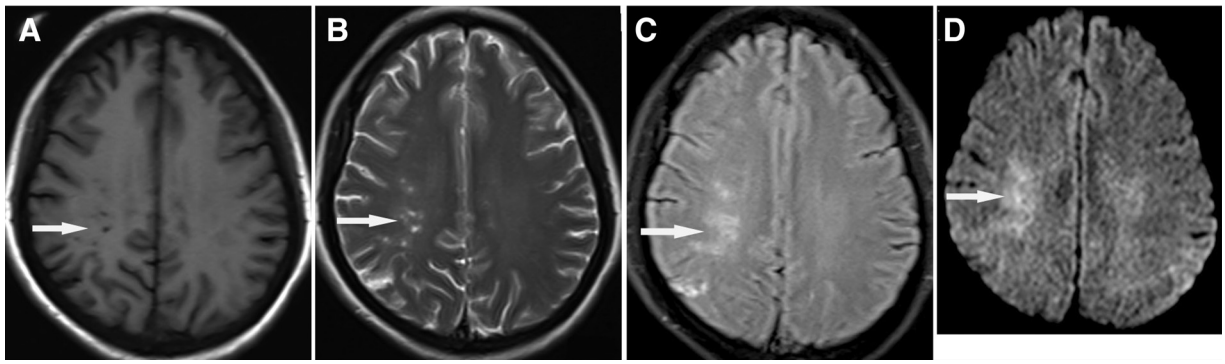

Figure 1. *MRI findings demonstrated cerebral infarction in centrum semiovale. (A and B) Axial T1-weighted image showing marked multiple hypointensity (white arrow) and T2-weighted image showing marked multiple hyperintensity (white arrow) in centrum semiovale. (C and D) Fluid-attenuated inversion recovery sequence and diffusion-weighted imaging, axial MRI demonstrating hyperintensity (white arrow) in centrum semiovale.*

mild mitral valve regurgitation and the left ventricular ejection fraction is 67%. Color Doppler flow imaging examination showed a mild resistance index of the intracranial segment of the vertebral artery (VA) and BA. Brain axial T2-weighted and diffusion-weighted imaging magnetic resonance imaging showed an area of hyperintensity in the right centrum semiovale, indicating acute cerebral infarction, consistent with his weakness in the left upper extremity (Fig 1). 3D time of flight magnetic resonance angiography (3D-TOF MRA) on 1.5 Tesla demonstrated the PPTA between right ICA and distal portion of BA and hypoplasia of the distal ICA (Fig 2).

Discussion

Blood vessel development in different embryological stages has been thoroughly reviewed.[2,7] PPTA is the most common vestigial artery observed and account for 85% in all observed persistent primitive artery.[2,7] The incidence of PPTA was reported to be .03%-2.2% of cerebral angiograms.[8,9] PPTA is female sex predilection, and it may be founded in patients of any age, on either side, and, in association with various vascular variants.[1]

PPTA *ordinarily arises from the cavernous segment of ICA and anastomosis in the mid or distal of the BA, closely runs along the trigeminal nerve*. PPTA usually supplies blood to both posterior cerebral arteries (PCAs) and superior cerebellar arteries (SCAs) via the distal BA. There have no flow-related stimulus for the BA proximal to the anastomosis to develop along with the embryo when PPTA persists. Thus might result in the frequent association of BA hypoplasia with PPTA. PPTA was divided into 2 types: medial (sphenoid type, hereafter referred to as medial type) or lateral type (petrosal type, hereafter referred to as lateral type). The medial PPTA runs out of the medial wall of the cavernous sinus and into the sella, thereafter perforates the clivus and end with the BA.[2,10,11] Lateral PPTA runs out of petrous part of temporal bone from the lateral wall of the cavernous sinus and from outside to inside arc anastomosis with the BA.[2,10,11] According to the configuration of ipsilateral PCA, PPTA is also separated into 2 types: Saltzman type I and Saltzman type II.[12] In addition, variant type PPTA, also known as type Ⅲ a, Ⅲ b, Ⅲ c, which the PPTA anastomose with superior cerebellar artery , anterior inferior cerebellar artery , and posterior inferior cerebellar artery, respectively.[12-15] So, MRA findings in our patient indicated the PPTA was type I (lateral type) (Fig 2).

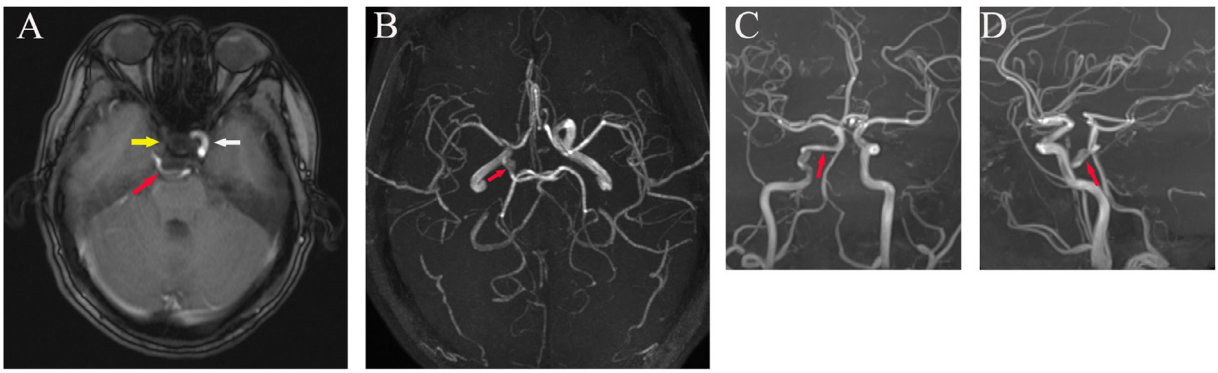

Figure 2. *MRA images demonstrating right PPTA and hypoplasia of internal carotid artery distal anastomosis. (A) 3D-TOF-MRA indicated that lateral type PPTA (red arrow) arc run toward ventral pons and left ICA (red arrow) in the right cavernous sinus segment. It is also indicating cavernous sinus segment of right ICA is absent (yellow arrow). (B, C, D) MRA demonstrating right PPTA (red arrow), right ICA distal anastomosis and bilateral PCoAs are absent (red arrow), hypoplasia in proximal BA anastomosis and bilateral VAs (red arrow). (Color version of figure is available online.)*

PPTA generally lacks specific clinical manifestation, but it can result in some secondary harms: (1) compress neighboring vessel and nerve. The lateral type PPTA can result in trigeminal neuralgia,[16] oculomotor palsy,[17] blindness,[18] tinnitus.[19] The medial type PPTA compress the pituitary gland and stalk and elevate blood prolactin level.[20] (2) Compress ventral brainstem and lead to pyramidal tract lesion. (3) Affect the normal pattern of brain blood flows. Thrombosis, stenosis, and dissection of ICA coexist with PPTA can lead to posterior circulation infarction.[21,22] Some studies have indicated that approximately 75% of PPTA coexist with simultaneous dysplasia in proximal BA anastomosis and bilateral VA, but the exact mechanism resulting in PPTA and hypoplasia in proximal BA and bilateral VA is not clear.[12,23]

The patient was 48 years old at the time of onset, so we executed the related risk factors screening for young adult stroke,[24–26] such as protein-C/S, NT-pro-BNP, lupus antibody, anticardiolipin antibodies, 24-hour Holter electrocardiography, echocardiography, and so on. Related risk factors for young adult stroke in our patient are normal. In our case, MRA findings indicated that the right ICA distal anastomosis was not developed. Corresponding MCA and ACA are supplied by contralateral ICA through anterior cerebral artery. The MCA was intact, but the subcortical distal branch may be insufficiency. So we speculated when the hypoplasia of proximal BA, bilateral VA, and PCoA coexisted with insufficiency of subcortical branch, PCA is also hypoperfusion, which finally led to cerebral watershed infarction (posterior cortical type) in the right parietal lobe (Fig 1).

Conclusions

PPTA occasionally company with stenosis, occlusion, dissection, and congenital absence of ICA, but all they are happened in proximal anastomosis of PPTA.[27,28] As far as we know, right PPTA and the homolateral hypoplasia of ICA distal anastomosis are not reported. PPTA usually coexists with many vascular variants and lacks special clinical manifestations. Finally, the extremely rare MR findings were shown in our case. It is not clear whether the PPTA is associated with ischemic cerebrovascular disease. Early identification of PPTA and its related mutations will be conducive to provide further treatment strategies.

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Availability of Data and Materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Authors' Contributions

L.D.P. carried out data collection and drafted the manuscript. W.Y. and Z.Z.H. conceived, designed the research and interpreted the data and made critical revisions of the manuscript. W.K., Y.M. and W.Y.F. participated in data collection and interpreted the data. All authors read and approved the final manuscripts.

Ethics Approval and Consent to Participate

The principles of the Declaration of Helsinki were followed and this study was approved by the ethics committee of the Fourth Affiliated Hospital of Anhui Medical University.

Consent for Publication

Written informed consent was obtained from the participant.

Competing Interests

All the authors declare that they have no competing interests.

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