Bilateral persistent primitive hypoglossal arteries associated with unilateral symptomatic carotid thromboembolism

Riddhi Patira[1*], Christopher Kyper[2], Pallav Shah[3], Kadir Erkmen[4]

1. Department of Neurology, Temple University Hospital, Philadelphia, USA

2. Lewis Katz School of Medicine at Temple University, Philadelphia, USA

3. Department of Radiology, Temple University Hospital, Philadelphia, USA

4. Department of Neurosurgery, Temple University Hospital, Philadelphia, USA

* **Correspondence:** Riddhi Patira, 1420 Locust St., Apt. 23J, Philadelphia, 19102, PA, USA (*patiriddhi@gmail.com*)

Radiology Case. 2017 Apr; 11(4):1-9 :: DOI: 10.3941/jrcr.v11i4.3010

ABSTRACT

We report the fifth case of bilateral persistent primitive hypoglossal arteries in the literature. This is also the first such case to be demonstrated on computerized tomography angiogram (CTA) and the first case to be associated with a symptomatic carotid thrombus. The sub-occlusive thrombus was distal to the take-off of the dominant persistent hypoglossal artery (PHA) from the internal carotid artery, thus sparing involvement of posterior circulation. Timely identification of the internal carotid artery thromboembolism in the setting of a PHA is important to allow for thrombectomy. Any intervention was not done in this case as the patient was out of the window and at an additional risk of inducing intracranial thromboembolism. Symptomatic carotid stenosis at the PHA take-off is typically treated with endovascular angioplasty and stenting due to the typically high level of the bifurcation in the neck.

CASE REPORT

CASE REPORT

A 79-year-old right-handed man with no significant past medical history presented with wake-up symptoms of left-sided hemiplegia, left homonymous hemianopia, left-sided visual-spatial neglect, and profound anosognosia. The initial score on the NIH (National Institutes of Health) stroke scale was 22. The patient was last known to be normal 13 hours prior to arrival, and was thus ineligible for both intravenous and endovascular thrombolysis. Initial brain computerized tomography (CT) did not show any signs of ischemia. A computerized tomography angiogram (CTA) of the head and neck demonstrated that the vertebrobasilar circulation was filled through bilateral fetal persistent hypoglossal arteries (PHAs) originating from the respective internal carotid arteries (ICA). The right PHA was dominant and coursed superiorly by entering the intracranial space through the right hypoglossal canal to form the basilar artery, thereby becoming the major vessel feeding the posterior circulation (Figure 1). The left PHA was hypoplastic, and entered the intracranial space through the left hypoglossal canal, eventually terminating into the ipsilateral posterior inferior cerebellar artery (PICA) (Figure 2). A sub-occlusive filling defect was visualized within the right ICA just distal to the origin of the right dominant PHA, consistent with an intraluminal thrombus measuring 12 mm X 3 mm (pink arrow in Figure 1). There was also a region of mild calcification in right ICA proximal to the thrombus without any significant stenosis (red arrows in Figure 3a and

3b). The thrombus had no extension into the PHA (yellow arrow in Figure 1a), thus sparing the posterior circulation (Figure 4c and 4d). Both vertebral arteries were absent (Figure 4e and 4f). A computerized tomography perfusion (CTP) was performed along with CTA which showed an area of ischemic penumbra within the right posterior middle cerebral artery (MCA) distribution and core infarction in the right anterior MCA distribution. Despite the presence of a viable penumbra, mechanical thrombectomy was not performed for multiple reasons: the patient arrived outside the appropriate window for the thrombectomy, there was possibility of converting a cervical thromboembolism to an intracranial thromboembolism, and there was additional risk of embolization to the posterior circulation through the patent PHA. The patient was medically optimized with permissive hypertension, and was started on a heparin drip due to the sub-occlusive nature of the ICA thrombus to prevent complete occlusion or further propagation. A subsequent brain magnetic resonance imaging (MRI) on the third day of hospitalization demonstrated extensive infarction of the right cerebral hemisphere in the territory of the right MCA (Figure 4a-b), with no ischemia in areas supplied by posterior circulation (Figure 4c-d). Magnetic resonance angiogram (MRA) showed a patent right MCA and a patent basilar artery (Figure 5). The next day, the patient's exam worsened and a repeat non-contrast brain CT showed hemorrhagic conversion of the infarct with mass effect, prompting discontinuation of heparin. No surgical intervention was necessary for midline shift due to clinical stability of the patient. The remainder of the patient's hospital course was uneventful and he demonstrated minimal recovery upon discharge. A repeat CTA was planned 4-6 weeks after discharge; however, the patient passed away at home from respiratory distress. An autopsy was not performed.

DISCUSSION

Etiology & Demographics:

Unilateral persistent hypoglossal artery (PHA) is the second most common persistent fetal carotid-vertebrobasilar anastomosis after trigeminal artery, with a reported prevalence of 0.29% [1]. In early stages of embryonic development, there are four types of fetal carotid-vertebrobasilar anastomoses which include the primitive trigeminal, hypoglossal, otic (acoustic), and proatlantal intersegmental arteries (Figure 6). As the embryo develops, the posterior communicating arteries develop and the anastomotic arteries begin to regress at approximately the 30th to 40th day of fetal development. Failure of regression of these primitive anastomoses leads to their persistence in the fetus and into adulthood. As a result of persistence of the PHA, the vertebral arteries are either hypoplastic or altogether absent. PHA can be differentiated from other anastomotic variants by its origin from the cervical carotid between C1-C3 vertebral levels and its entry into the intracranial space through the hypoglossal canal. While passing through the hypoglossal canal, the PHA can cause compression of the hypoglossal nerve leading to hypoglossal neuralgia or paresis, which was not reported by our patient. The classification scheme of unilateral PHA into two types based on its origin either from the ICA (Type 1) or the external carotid artery (Type 2) [1] does not apply to bilateral PHA as these always arise from ICA. In a large cohort of CTA and MRA, none of the cases demonstrated bilateral PHA, making bilateral PHA an extremely rare fetal variant [1]. Only four cases of bilateral PHA have been reported in literature.

Clinical & Imaging findings:

Three cases of bilateral PHA were identified with conventional angiogram, all of which were associated with intracranial hemorrhage. The first case had no clear pathology [2]. The other two had an aneurysm at the junction of the PHA and the basilar artery- in one case, this was successfully clipped [3]; and in the other case, it was not attributed as the cause of perimesencephalic sub-arachnoid hemorrhage [4]. The fourth case of bilateral PHA was discovered on MRA when a routine carotid ultrasound performed by a primary care doctor for general health screening showed incidental absence of flow velocities in the vertebral arteries. This prompted the MRA, which showed no associated pathology [5]. We report the fifth case of bilateral PHA and the first case to be demonstrated on CTA. Furthermore, bilateral PHA have never been reported with carotid stenosis or occlusion, and this is also the first case of bilateral PHA to be associated with a symptomatic carotid thromboembolism.

Differential Diagnoses:

As we have learned from the cases of unilateral PHA, if stenosis is present proximal to the origin of the PHA, simultaneous infarctions may occur in both the anterior and posterior circulation mimicking a cardioembolic source; and if completely occluded, infarction in the ipsilateral hemisphere and entire brainstem can result in brain death due to herniation [6]. Thus, when the source of emboli is unclear, vascular imaging may demonstrate fetal variants connecting anterior and posterior circulation that can cause emboli to multiple vascular territories. An aortic pathology can also cause vessel-to-vessel embolization in multiple vascular territories which can be diagnosed with a trans-esophageal echocardiogram.

Treatment & Prognosis:

Timely identification of ICA occlusion with PHA could allow for mechanical thrombectomy, which was not done in this case for reasons aforementioned. The origin of the PHA is usually from the ICA at the C1-C3 vertebral level. Any significant carotid stenosis is generally treated with endovascular stenting with distal embolic protection due to its origin at a high vertebral level [7-10]. In one case, successful revascularization was achieved by endarterectomy with shunt positioning based upon extension of the plaque into the PHA [11]. In our case, the origin of the PHA was at the level of C1 (Figure 1). If our patient had presented with transient or non-disabling symptoms with proximal ICA stenosis, ICA balloon angioplasty and stenting would have been a viable option for revascularization.

TEACHING POINT

Fetal anastomotic variants can create misleading clinical findings in the patients presenting with stroke syndromes. Arterial emboli originating from a fetal anastomotic vessel can result in stroke that can mimic a cardio-embolic source due to ischemia in multiple vascular territories typically supplied by separate vessels. Timely identification of pathology including atherosclerotic plaque associated with these vascular variants on non-invasive imaging like computerized tomography angiogram is critical to identify the source of stroke and in guidance of potential therapy including surgical or endovascular intervention.

REFERENCES

1. Uchino A, Saito N, Okada Y, Kozawa E, Nishi N, Mizukoshi W, Inoue K, Nakajima R, Takahashi M. Persistent hypoglossal artery and its variants diagnosed by CT and MR angiography. Neuroradiology. 2013 Jan 1;55(1):17-23. PMID: 22821359.

2. Karasawa J, Kikuchi HA, Furuse SE, Sakaki TO, Yoshida YA, Ohnishi HI. Bilateral persistent carotid-basilar anastomoses. American Journal of Roentgenology. 1976 Dec 1;127(6):1053-6. PMID: 998821.

3. Murayama Y, Fujimoto N, Matsumoto K. Bilateral persistent primitive hypoglossal arteries associated with a large ruptured aneurysm on one side. Surgical neurology. 1985 Nov 30;24(5):498-502. PMID: 4049224.

4. Garge S, Moses V, Keshava S, Ahmed M, Moorthy R. Persistent hypoglossal arteries with aneurysmal dilation of left hypoglossal artery: a rare case report and review of the literature. BJR case reports. 2016 May:20150301.

5. Takahashi H, Tanaka H, Fujita N, Tomiyama N. Bilateral persistent hypoglossal arteries: MRI findings. The British journal of radiology. 2014 Jan 28. PMID: 3473955.

6. Kawano H, Inatomi Y, Hirano T, Yonehara T. Cerebral infarction in both carotid and vertebrobasilar territories associated with a persistent primitive hypoglossal artery with severe dilated cardiomyopathy. Journal of Stroke and Cerebrovascular Diseases. 2014 Jan 31;23(1):176-8. PMID: 22959108.

7. Zhang L, Song G, Chen L, Jiao L, Chen Y, Wang Y. Concomitant asymptomatic internal carotid artery and persistent primitive hypoglossal artery stenosis treated by endovascular stenting with proximal embolic protection. Journal of vascular surgery. 2016 Jan 31;63(1):237-40. PMID: 24877853.

8. Silva CF, Hou SY, Kühn AL, Whitten RH, Wakhloo AK. Double embolic protection during carotid artery stenting with persistent hypoglossal artery. Journal of neurointerventional surgery. 2014 Apr 1;6(3):e23-. PMID: 23645663.

9. Murai S, Kusaka N, Umakoshi M, Itami H, Otsuka S, Nishiura T, Ogihara K. Stenting for Internal Carotid Artery Stenosis Associated with Persistent Primitive Hypoglossal Artery Using Proximal Flow Blockade and Distal Protection System: A Technical Case Report and Literature Review. Journal of Stroke and Cerebrovascular Diseases. 2016 Jun 30;25(6):e98-102. PMID: 27105567.

10. Ryu B, Ishikawa T, Hashimoto K, Shimizu M, Yagi S, Shimizu T, Kawamata T. Internal carotid artery stenosis with persistent primitive hypoglossal artery treated with carotid artery stenting: A case report and literature review. The neuroradiology journal. 2016 Apr 1;29(2):115-21. PMID: 26825135.

11. Koleilat I, Hanover T. Carotid Endarterectomy in the Face of a Persistent Hypoglossal Artery. Annals of vascular surgery. 2015 Nov 30;29(8):1660-e1. PMID: 26303270.

FIGURES

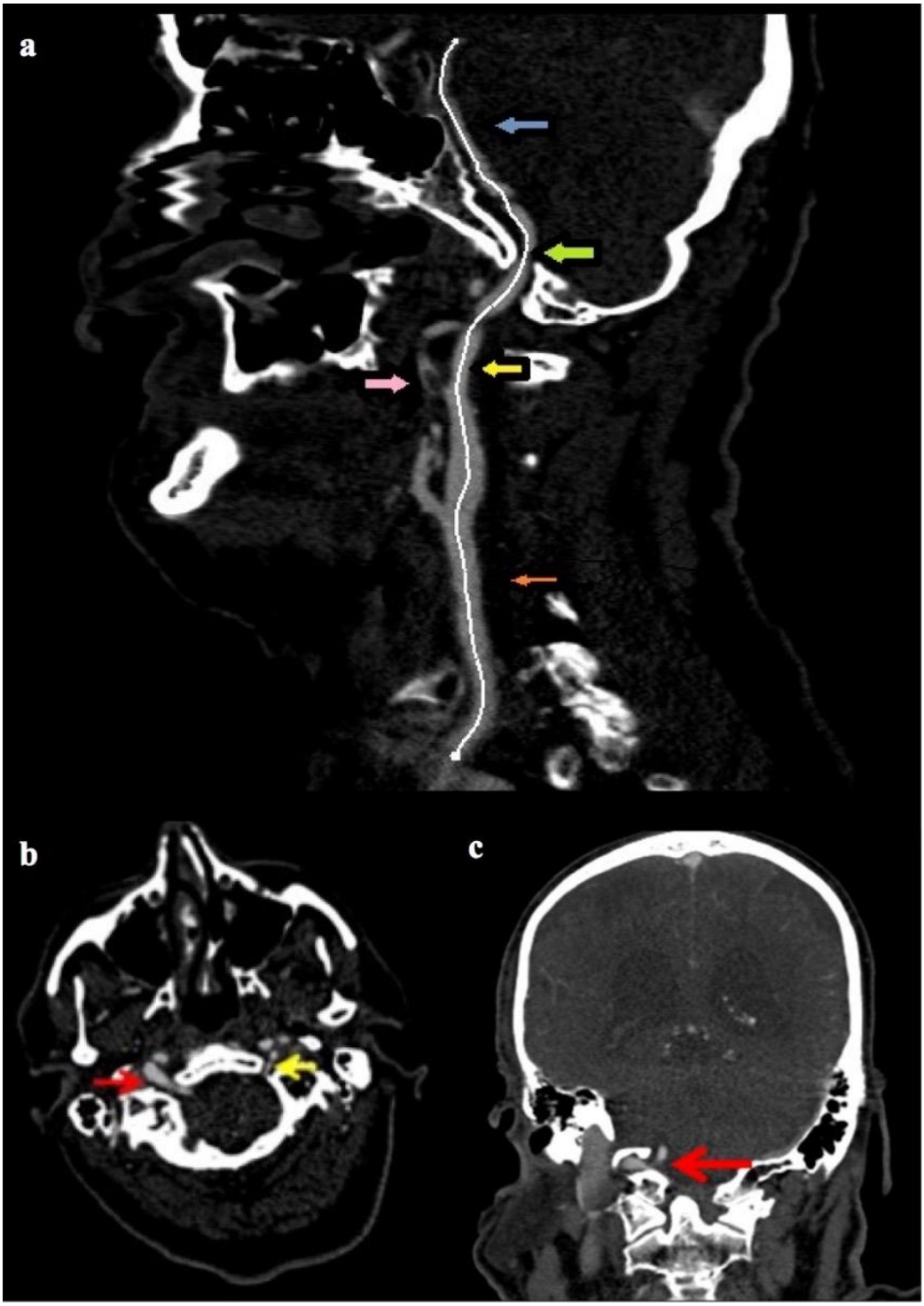

Figure 1: A 79 year-old man with a right middle carotid artery territory infarct secondary to sub-occlusive thrombus within the right internal carotid artery (ICA) in the setting of bilateral persistent hypoglossal arteries (PHAs).
Findings:
(a) Sagital view of the CTA with vessel-tracking by Tera Recon (white line). The right common carotid artery (inferior aspect of white line depicted by an orange arrow) courses superiorly to branch into the external carotid artery and the ICA. The right ICA continues (white line) until it gives off a right PHA (segment of white line pointed by yellow arrow) and remainder of ICA is further shown to have a sub-occlusive filling defect just distal to the origin of the right PHA, consistent with an intraluminal thrombus (pink arrow). The right PHA (continues as white line pointed by yellow arrow) enters the intracranial space through the right hypoglossal canal (green arrow) and terminates into the basilar artery (blue arrow).
(b) Axial view of the CTA and (c) Coronal view of the CTA depicting a large right PHA (red arrow) traversing the hypoglossal canal into the intracranial space, and a smaller left PHA (yellow arrow) entering the left hypoglossal canal.
Technique: mA 116, KVp 120, slice thickness 0.60 mm, with 80 mL of Omnipaque 350

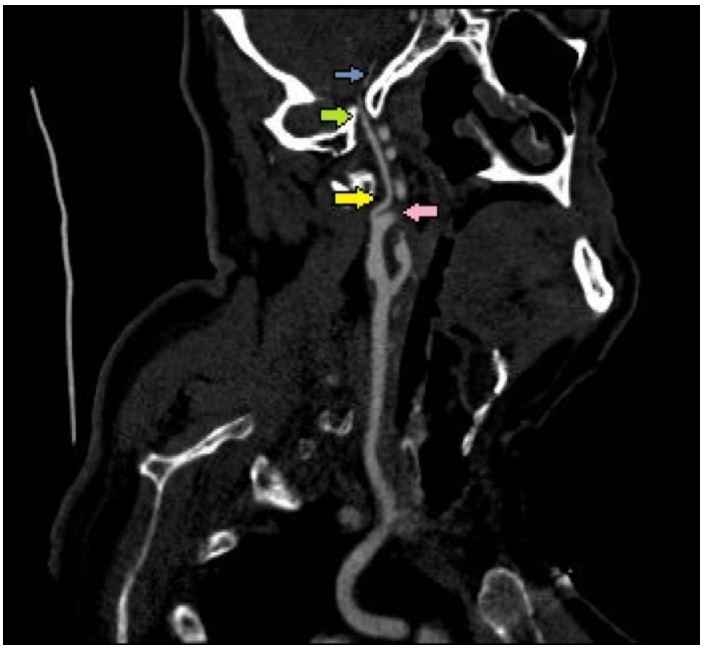

Figure 2: A 79 year-old man with a right middle carotid artery territory infarct secondary to sub-occlusive thrombus within the right internal carotid artery (ICA) in the setting of bilateral persistent hypoglossal arteries (PHAs).
Findings: Sagital view of CTA with vessel-tracking showing the left PHA (yellow arrow) originating from the left ICA (pink arrow), entering the intracranial space through the left hypoglossal canal (green arrow) to terminate into the ipsilateral posterior inferior cerebellar artery (blue arrow).
Technique: mA 116, KVp 120, slice thickness 0.60 mm, with 80 mL of Omnipaque 350

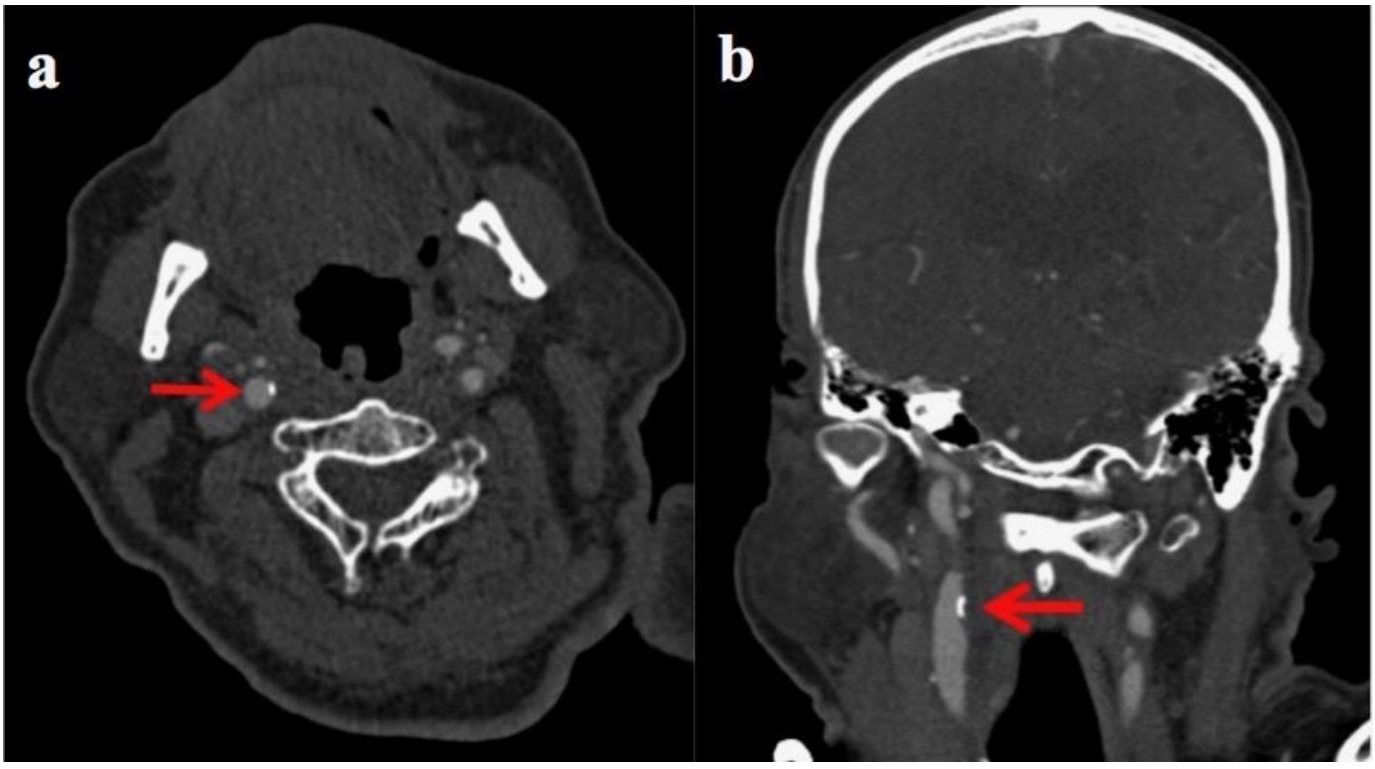

Figure 3: A 79 year-old man with a right middle carotid artery territory infarct secondary to sub-occlusive thrombus within the right internal carotid artery (ICA) in the setting of bilateral persistent hypoglossal arteries (PHAs).
Findings: (a) Axial view and (b) Coronal view of CTA depicting a calcified plaque (both red arrows) proximal to the suspected thrombus without any significant stenosis at the level of the plaque.
Technique: mA 239, KVp 120, slice thickness 1.00 mm, with 80 mL of Omnipaque 350

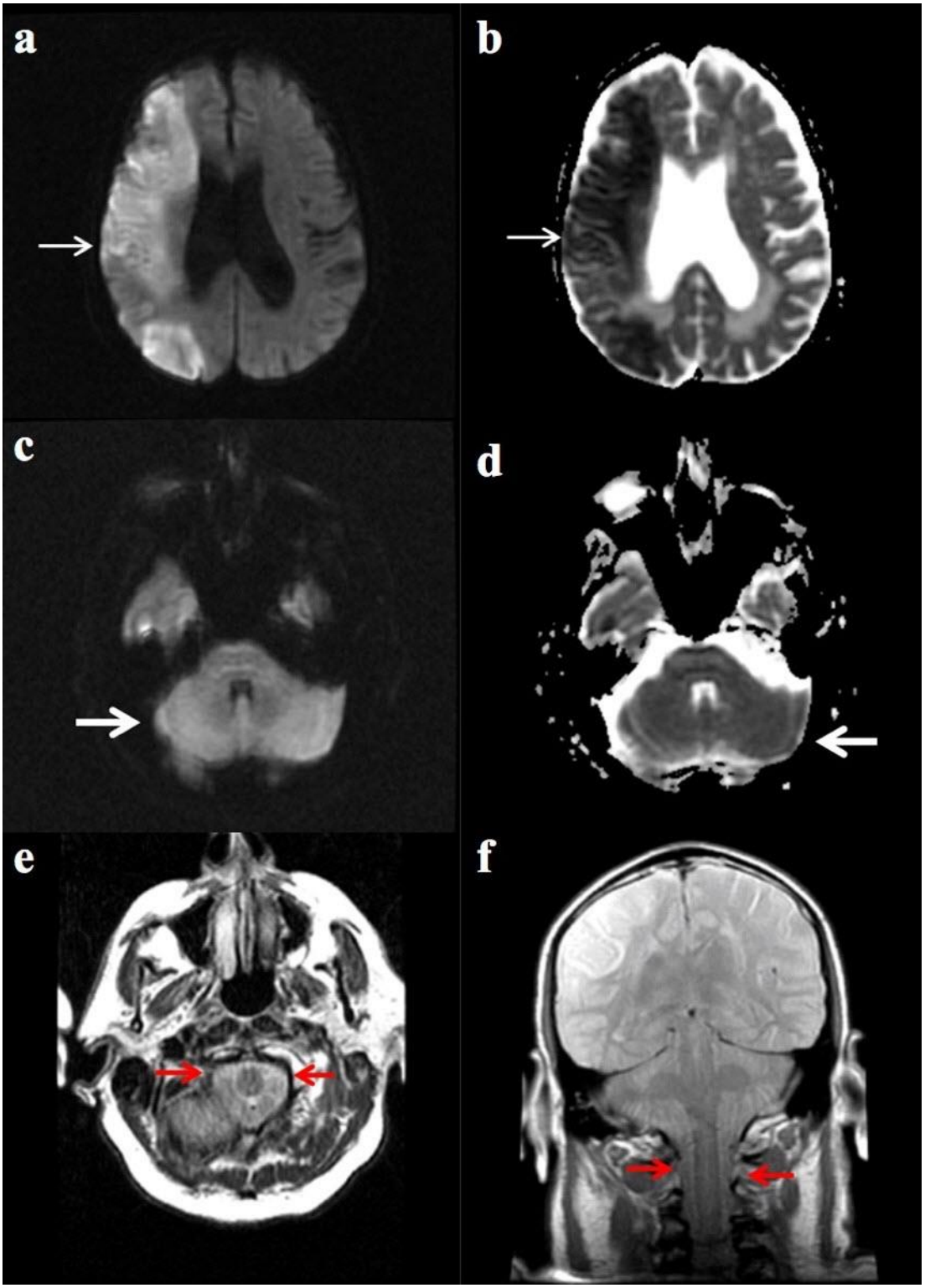

Figure 4: A 79 year-old man with a right middle carotid artery territory infarct secondary to sub-occlusive thrombus within the right internal carotid artery (ICA) in the setting of bilateral persistent hypoglossal arteries (PHAs).
Findings: (a) Axial diffusion-weighted (DWI) sequence showing a large area of restricted diffusion (white arrow) with (b) matched defect in corresponding ADC map within the territory supplied by the right middle cerebral artery suggesting a right MCA infarct (white arrow). (c) Axial DWI and (d) corresponding ADC map showing cerebellum and brainstem without any areas of restricted diffusion (both white solid arrows). Rest of the areas supplied by the posterior circulation were also spared (not shown here). (e) Axial T2 and (f) coronal proton density sequence showing absence of bilateral vertebral arteries (both red arrows).
Technique:
(a) - (d): 1.5 T GE Magnet, TR: 9000, TE: 78.8, FOV: 23 cm. (e): 1.5 T GE Magnet, TR: 2516.66, TE: 82.8, FOV: 23 cm. (f): 1.5 T GE Magnet, TR: 2500, TE: 19.66, FOV: 23 cm

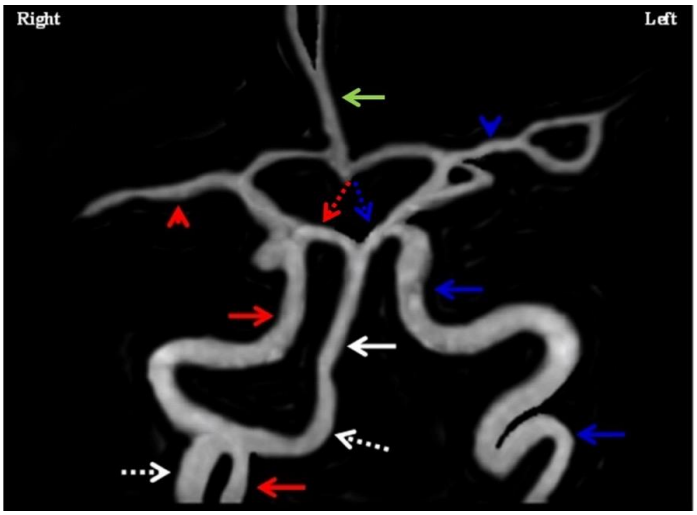

Figure 5: A 79 year-old man with a right middle carotid artery territory (MCA) infarct secondary to sub-occlusive thrombus within the right internal carotid artery (ICA) in the setting of bilateral persistent hypoglossal arteries (PHAs).
Findings: MRA showing patent basilar artery (white solid arrow) getting its major supply from right PHA (white dotted arrows). Right MCA is patent (red arrowhead). Other patent major intracranial vessels are shown including right ICA (red arrows), left ICA (blue arrows), left MCA (blue arrowhead), right posterior cerebral artery (red dotted arrow), left posterior cerebral artery (blue dotted arrow), and anterior cerebral artery (green solid arrow).
Technique: 1.5 T GE Magnet, TR 25, TE: 2.4.

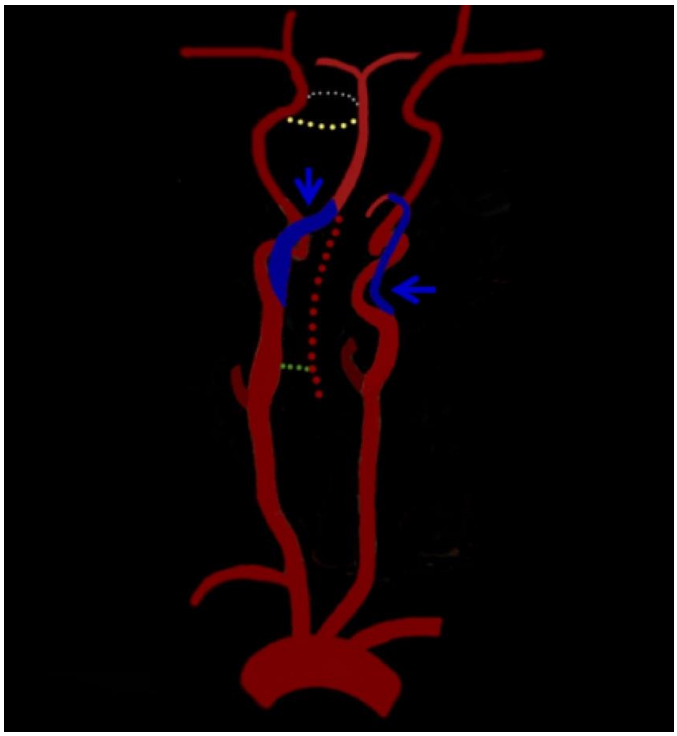

Figure 6: A 79 year-old man with a right middle carotid artery territory infarct secondary to sub-occlusive thrombus within the right internal carotid artery (ICA) in the setting of bilateral persistent hypoglossal arteries (PHAs).
Findings: Anatomic illustration showing both PHA (blue segments pointed by blue arrows) originating from respective ICA. In addition, though not present in our case, the location of other fetal variants are shown. The persistent trigeminal artery (white dotted line) is an anastomosis between the distal part of ICA and the distal part of the basilar artery. The persistent otic artery (yellow dotted line) is an anastomosis between the ICA and the proximal part of the basilar artery. The persistent postatlantal artery (green dotted line) is an anastomosis between the proximal part of ICA and the ipsilateral vertebral artery (red dotted line).

Diagnosis	MRI	MRA	CTA	Ultrasound
Carotid stenosis or occlusion proximal to the PHA	Diffusion-weighted imaging may show areas of restricted diffusion consistent with acute infarction in the anterior circulation on side of carotid stenosis/occlusion as well as in the posterior circulation.	• Time-of-flight MRA shows PHA originating from the respective ICA at the C1-C3 vertebra level. • Constructive interference in steady state (CISS) can identify the PHA by demonstrating its entry into intracranial space through the hypoglossal canal. • The stenosis or occlusion is present proximal to the origin of the PHA. • Vertebral artery is either absent or hypoplastic on the side of PHA	• PHA usually originates from the respective ICA at the C1-C3 vertebra level. • Axial view of CTA can help identify the PHA by demonstrating its entry into intracranial space through the hypoglossal canal. • The stenosis or occlusion is present proximal to the origin of the PHA. • Vertebral artery is either absent or hypoplastic on the side of PHA.	• Increased peak systolic flow velocity may be seen in the region of carotid stenosis. • Absence of flow velocity may be seen in the respective vertebral artery due to its absence.
Carotid stenosis or occlusion distal to the PHA	• Diffusion-weighted imaging may show areas of restricted diffusion consistent with acute infarction in anterior circulation on side of carotid stenosis/occlusion. • The posterior circulation would be spared with no areas of restricted diffusion.	• MRA shows PHA originating from the respective ICA at the C1-C3 vertebra level. • Constructive interference in steady state (CISS) identify the PHA by demonstrating its entry into intracranial space through the hypoglossal canal. • The stenosis or occlusion is present distal to the origin of the PHA. • Vertebral artery is either absent or hypoplastic on the side of PHA	• PHA usually originates from the respective ICA at the C1-C3 vertebra level. • Axial view of CTA can identify the PHA by demonstrating its entry into intracranial space through the hypoglossal canal. • The stenosis or occlusion is present distal to the origin of the PHA. • Vertebral artery is either absent or hypoplastic on the side of PHA	• Increased peak systolic flow velocity may be seen in the region of carotid stenosis. • Absence of flow velocity may be seen in the respective vertebral artery due to its absence.
Carotid stenosis/ occlusion in absence of anomalies like PHA	• Diffusion- weighted imaging may show areas of restricted diffusion consistent with acute infarction in anterior circulation on side of carotid stenosis/occlusion. • The posterior circulation would be spared with no areas of restricted diffusion.	• Absence of anomaly like PHA on time-of-flight MRA. • Carotid stenosis is present on the side of the infarction on time-of-flight MRA. • Both vertebral arteries will be present. One of the vertebral arteries may be hypoplastic in presence of other being dominant.	• Absence of anomaly like PHA. • Carotid stenosis is present on the side of the infarction. • Both vertebral arteries will be present. One of the vertebral arteries may be hypoplastic in presence of other being dominant.	• Increased peak systolic flow velocity may be seen in the region of carotid stenosis. • Presence of flow velocities seen in both vertebral arteries.
Cardiac source of embolization	Diffusion-weighted imaging may show areas of restricted diffusion consistent with acute infarction occurs on both sides of the anterior circulation as well as in the posterior circulation.	• Absence of anomaly like PHA on time-of-flight MRA. • Carotid stenosis/occlusion may not occur. • Both vertebral arteries will be present. One of the vertebral arteries may be hypoplastic in presence of other being the dominant.	• Absence of anomaly like PHA. • Carotid stenosis/occlusion may not occur. • Both vertebral arteries will be present. One of the vertebral arteries may be hypoplastic in presence of other being dominant.	• The flow velocities may be seen normal in both the carotid and vertebral arteries in absence of any pathology.

Table 1: Differential diagnosis for right hemispheric infarct.

Etiology	Persistent fetal carotid-vertebrobasilar anastomosis
Incidence	Only 4 cases are reported in literature. This is fifth case of bilateral persistent hypoglossal artery (PHA) and first case of bilateral PHA to be reported on computerized tomography angiogram (CTA).
Gender ratio	M : F = 1 : 1
Age predilection	30s – 70s
Risk factors	Developmental anomaly
Treatment	• If associated with carotid stenosis, endovascular angioplasty with stenting is the treatment of choice. • If associated with aneurysm, surgical clipping is an option. • If incidentally discovered with no associated pathology, no treatment is required.
Prognosis	• In associated with carotid stenosis, both endovascular stenting and endarterectomy have been successfully performed with resultant reperfusion. • If associated with ruptured aneurysm, surgical clipping is successful.
Findings on imaging	• PHA originates at the C1-C3 vertebra level from the respective internal carotid artery (ICA) and enters the intracranial space through the respective hypoglossal canal. • Vertebral artery is either absent or hypoplastic on the side of PHA. • Aneurysms may occur at the junction of the PHA and the basilar artery. • Carotid stenosis may occur with or without the involvement of PHA. • PHA may be incidental with no associated vascular pathology.

Table 2: Summary table for characteristics of bilateral PHA.

ABBREVIATIONS

CT = computerized tomography
CTA = computerized tomography angiogram
CTP = computerized tomography perfusion
ICA = internal carotid artery
MRA = magnetic resonance angiogram
MRI = magnetic resonance imaging
PHA = persistent hypoglossal artery
PICA = posterior inferior cerebellar artery

KEYWORDS

Bilateral persistent hypoglossal artery; carotid thromboembolism; computed tomography angiogram; endovascular stenting; fetal variant

Online access

This publication is online available at:
www.radiologycases.com/index.php/radiologycases/article/view/3010

Peer discussion

Discuss this manuscript in our protected discussion forum at:
www.radiolopolis.com/forums/JRCR

Interactivity

This publication is available as an interactive article with scroll, window/level, magnify and more features.
Available online at www.RadiologyCases.com

Published by EduRad

www.EduRad.org