Variation of a Persistent Primitive Hypoglossal Artery (PPHA) as Incidental Finding in the Diagnostic Clarification of Cerebral Vasculopathy Associated with Intracranial Vasculitis

S. Hopf-Jensen, MD¹ L. Marques, MD¹ M. Preiß, MD¹ W. Börm, MD, PhD²
S. Müller-Hülsbeck, MD, PhD¹

¹ Department of Diagnostic and Interventional Radiology and
Neuroradiology, Diakonissenhospital Flensburg, Flensburg, Germany

² Department of Neurosurgery, Diakonissenhospital, Flensburg, Germany

Address for correspondence Silke Hopf-Jensen, MD, Knuthstraße 1,
Department of Diagnostic and Interventional Radiology and
Neuroradiology, Diakonissenhospital Flensburg, Flensburg, Germany
(e-mail: hopfsi@diako.de).

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Abstract

We present a very rare variation of a persistent primitive hypoglossal artery (PPHA) arising from the internal carotid artery, detected during a diagnostic angiography. A 50-year-old female patient was admitted with an atypical intracranial hematoma in the left frontal lobe. Catheter angiography revealed intracranial vasculopathy with segmental stenoses, a small aneurysm of the right internal carotid artery bifurcation and a “string of beads” appearance of the left carotid artery, consistent with fibromuscular disease. On the left side, a vertebral artery ending in the posterior inferior cerebellar artery (PICA) was detected, whereas on the right side the vertebral artery was aplastic. During selective angiography of the right common carotid artery, a persistent hypoglossal artery was seen supplying the basilar artery. The literature of persistent embryonal carotid-vertebrobasilar anastomosis and their anatomical variations is discussed with respect to clinical importance for ischemia, interventional procedures, and surgery.

Keywords

- ▶ aneurysm
- ▶ artery
- ▶ vasculitis
- ▶ vascular access
- ▶ symptomatic
- ▶ internal carotid artery surgery.

Persistent primitive hypoglossal artery (PPHA) is a type of carotid-basilar anastomosis with an incidence of 0.027 to 0.26%.^{1–3} This congenital vessel courses through an enlarged anterior condyloid foramen (hypoglossal canal) and joins the basilar artery often in addition to an ipsilateral aplastic vertebral artery and hypoplastic vertebral artery on the contralateral side, or hypoplastic vertebral arteries on both sides.³ Usually, carotid-vertebral and carotid-basilar anastomosis are asymptomatic incidental findings during angiography, but association with ischemic events, aneurysm,^{2,3} arteriovenous malformations,⁴ and nerve palsy has been described.^{1,2} A combination with vasculitis has not been reported in the literature before.

Case Report

A 50-year-old female patient was admitted to the hospital because of sudden onset of headache. No sensomotoric deficits were found in the neurological examination. The native computerized tomography (CT) and brain magnetic resonance imaging (MRI) revealed a cortical intracerebral hemorrhage in the left frontal lobe. Digital subtraction angiography showed the common carotid artery on the left side having a normal origin as second branch of the aortic arch. A filiform left vertebral artery ends in the posterior inferior cerebellar artery (PICA) following a regular extracranial course originating from the subclavian artery and entering

Fig. 1 Magnetic resonance angiography (coronal MIP time-of-flight) shows an absence of both vertebral arteries with the persistent primitive hypoglossal artery (PPHA, red arrow) following the extra-cranial course of the right internal carotid artery. The PPHA passes the widened hypoglossal canal joining the basilar artery.

the transverse foramina at the C4 level. No retrograde filling of the right vertebral artery was detected. The origin of the right vertebral artery from the brachiocephalic trunk was missing and a hypoplasia of this artery was assumed. During selective angiography of the right common artery, an ante-

Fig. 3 As additional finding vasculitis is detected (digital subtraction angiography [DSA] 90 degree lat. projection) magnified DSA (lat. projection 90 degrees) presents segmental stenoses (red arrow) affecting three terminal media branches typical for vasculitis.

grade filling of a PPHA was noted. This vessel arises from the right internal carotid artery proximal, ascends vertically, and then enters a widened hypoglossal canal to supply the basilar artery (► **Figs. 1** and **2**). In addition, a small saccular aneurysm located at the right internal carotid artery bifurcation was detected.

Intracranially, several segmental stenoses proximally and also in secondary and tertiary vessels were observed (► **Fig. 3**). After excluding numerous differential diagnoses, including Wegener granulomatosis and collagen vascular disorders in the setting of secondary central nervous system

Fig. 2 Digital subtraction angiography of the right internal carotid artery (ICA) and right common carotid artery demonstrates the persistent primitive hypoglossal artery (blue arrow) originating from ICA. (a) Right ICA, town projection cranial 28 degrees; (b and c) right common carotid artery: right anterior oblique -30 degrees, caudal -6, 9 degrees, (d) right common carotid artery: left anterior oblique 37 degrees, caudal -10 degrees; (e) posterior inferior cerebellar artery terminating left vertebral artery.

Fig. 4 Anatomic sketch depicts the right persistent primitive hypoglossal artery. rCCA: right common carotid artery, lCCA: left common carotid artery, rICA: right internal carotid artery, lICA: left internal carotid artery, rECA: right external carotid artery, and lECA: left external carotid artery.

(CNS) vasculitis, a primary angiitis of the central nervous system (PACNS) was assumed. To confirm the diagnosis biopsy is necessary, which was refused by the patient. Primarily, the patient underwent steroid therapy. Considering the patient's desire, the aneurysm was coiled with balloon assistance (► **Fig. 5**). The follow-up was unremarkable. After 6 months, no new intracranial hematoma was detected.

Discussion

Particularly with regard to the increasing number of interventional procedures in occlusions of proximal great vessels,

the knowledge of anatomic variations and collateral circulation of supra-aortal vessels is of utmost importance for better understanding the genesis of cerebrovascular disease. Normally, anomalies and variations in supra-aortal arteries origin are not symptomatic.⁵ Most variations in the origin of the supraaortic vessels are detected during anatomical cadaver studies.^{6–8}

According to persistent congenital carotid-vertebral and carotid-basilar anastomoses, the following arteries are described: proatlantal intersegmental artery, hypoglossal artery, otic artery, trigeminal artery, and posterior communicating artery. The last vessel is the only artery that normally persists, the other four usually regress completely.⁹ The persistent trigeminal artery can cause disorders of the 3rd, 4th, 5th, and 6th cranial nerves. The otic artery can lead to acoustic or facial nerves palsy. The PPHA can be associated with palsy of the 12th nerve, glossopharyngeal neuralgia or vascular disorder of the posterior circulation. The vertebral arteries are usually hypoplastic. They originate as branches of the cervical part of the internal carotid artery between the C1 and C3 levels, pass through the hypoglossal canal, and join the lower portion of the basilar artery.¹

In the literature, associations between cerebral infarction of carotid and vertebrobasilar territories and PPHA are described.^{10–12} The PPHA may serve as a pathway for an embolus originating from an atherosclerotic disease of the internal carotid artery to reach the posterior circulation.¹¹ Temporary clamping of the PPHA during carotid endarterectomy may increase the risk of ischemia.³ Yuasa et al postulated that cerebral blood flow in the ipsilateral internal carotid artery associated with a PPHA may be greater than the contralateral flow resulting in intimal injury and stenosis.¹³ Cardioembolic infarction could be associated with PPHA in the absence of stenotic internal carotid artery lesions, as a possible cause of multiple infarctions in both the carotid and vertebrobasilar territories.¹²

Fig. 5 Often as secondary finding in persistent primitive arteries a small sacular aneurysm at the carotid bifurcation is detected (a: digital subtraction angiography [DSA]). Follow-up angiography after balloon-assisted coiling (b). Selective DSA shows a small carotid-T-aneurysm (red arrow) before and after balloon assisted coiling, which is associated with persistent primitive hypoglossal artery.

Six cases of PPHAs originating from the external carotid artery have been reported, one was associated with subarachnoid hemorrhage.^{2,3} Intracranial aneurysms occur with a frequency of approximately 26% in the setting of PPHA.² Endovascular treatment of a PPHA aneurysm invaginating into the pontomedullary junction through an internal carotid artery-PPHA approach has been reported.^{14,15} In our case, the aneurysm was located at the bifurcation of the right internal carotid, indicating a weakness in the vessel's wall or hemodynamic stress in the setting of an assumed primary angiitis of the central nervous system (PACNS). The diagnosis of PACNS remains challenging. Cerebral arteriography, cerebrospinal fluid examination, and MRI alone or in combination do not have sufficiently demonstrated positive predictive value to establish a diagnosis. Histologic confirmation is required for the diagnosis of PACNS (► Figs. 4 and 5).¹⁶

Conclusion

This case describes a rare anatomical variation of an incidental PPHA joining the basilar artery in combination with an aplastic right vertebral artery and a left vertebral artery ending in PICA. The awareness of anatomical variations and persistent carotid-basilar anastomoses is of utmost importance for both interventional treatment and neurosurgery.

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