Unusual case of persistent primitive hypoglossal artery with anterior choroidal artery aneurysm in Chiari type I malformation

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Abstract
Persistent primitive hypoglossal artery (PPHA) is a rare form of persistent embryonic carotid-basilar anastomosis. We present an unusual case of PPHA and an anterior choroidal artery (AChoA) aneurysm associated with Chiari type I malformation. A 45-year-old woman presented with transient dizziness. Magnetic resonance imaging revealed Chiari type I malformation and a left AChoA aneurysm. Digital subtraction angiography incidentally revealed a left PPHA. To the best of our knowledge, this is the first reported case of Chiari malformation in conjunction with PPHA and aneurysms. In this case, the perfusion of the posterior circulation is completely dependent on PPHA. It is very important to identify such variant vessels and complex angioarchitecture before planning neuroendovascular or surgical intervention to prevent possible risks.

Key words: Aneurysm; anterior choroidal artery; Chiari malformation; persistent primitive hypoglossal artery

Introduction
Persistent primitive hypoglossal artery (PPHA) is a rare form of persistent embryonic carotid-basilar anastomosis. PPHA is sometimes associated with stenosis or posterior circulation aneurysms. Herein, we present an unusual case of PPHA and an anterior choroidal artery (AChoA) aneurysm associated with Chiari type I malformation.

Case History
A 45-year-old woman without noticeable past history presented with transient dizziness. There was no association between the symptom and her posture. Magnetic resonance imaging (MRI) revealed caudal displacement of the cerebellar tonsils 10 mm below the level of the foramen magnum and descent of the fourth ventricle below Twining’s line (which connects the anterior tuberculum sellae and the internal occipital protuberance). These findings led to the diagnosis of Chiari type I malformation without syringomyelia [Figure 1A and B]. The MRI also showed the left internal carotid artery (ICA) aneurysm. Subsequent digital subtraction angiography revealed a...
left PPHA, which arose from ICA at the level of C1-2, passed through a hypoglossal canal, and joined a basilar artery (BA) [Figure 2A-D]. The left vertebral artery (VA) from subclavian artery terminated at extracranial segment, whereas the right VA terminated as a posterior inferior cerebellar artery (PICA). The left PICA was branched from the left VA following the PPHA in the posterior fossa after passing through the hypoglossal canal. Posterior communicating arteries (PCoA) were not visualized bilaterally. Therefore, blood supply of the posterior circulation was dependent on PPHA [Figure 2E and F].

Selective left internal carotid angiography revealed an AChoA aneurysm approximately 4.5 mm in size, with a bleb at the anterolateral wall [Figure 3A-D]. The angle of the anterior knee of the carotid siphon was very sharp, causing the course of the ICA supraclinoid segment to be almost horizontal [Figure 3B and D]. Three-dimensional reconstruction imaging and additional computed tomography (CT) angiography showed that the aneurysm was located in a low position very close to a posterior clinoid process [Figure 3E and F]. Treatment strategies for the AChoA aneurysm have been under consideration, while comparing the risk of rupture and the risk of surgery with a focus on the above features.

Discussion

PPHA is the second most common persistent carotid-basilar anastomoses following persistent primitive trigeminal artery, and the incidence has been reported to be 0.03%–0.26%.[1] This vessel was first described by Batujeff in 1889,[2] and Lie proposed four criteria for the definition: 1) the PPHA arises from the ICA at the C1-3 level; 2) it enters the posterior cranial fossa via the hypoglossal canal; 3) the BA arises from a branch of PPHA; and 4) there is no ipsilateral PCoA.[3] Since these variations involved specific areas of the vascular tree, Lasjaunias and his colleagues proposed the concept of “segmental identity and vulnerability.”[4,5] Furthermore, there have been six reported cases of PPHA originating from the external carotid artery (ECA).[6] It is reasonable that PPHA also originates from the ECA, based on the hypothesis that it is a relic of ascending pharyngeal artery.[5] If a PPHA is present, 79%–90% are associated with bilaterally hypoplastic VAs or with a hypoplastic VA on one side and an absent VA on the other.[7-9] The contralateral VA and PCoA are only present in one-third of the cases. In such cases, the PPHA is the primary blood supply of the posterior circulation.

An association between PPHA and stroke due to stenosis or intracranial aneurysms has been noted. Past reviews reported that 26%–27.6% had aneurysms and that were located mainly (about 85%) in the posterior circulation, especially at the PPHA-BA junction.[7,10-12] Some of them caused cerebral infarction or subarachnoid hemorrhage and required surgical treatment. Recently, neuroendovascular therapy has tended to be selected, such as mechanical thrombectomy, carotid artery stenting, or intraneursymal...
coiling.\textsuperscript{[10]} As described above, occurrence of aneurysm in the anterior circulation region in PPHA is rare. In the present case, perfusion of the posterior circulation is completely dependent on PPHA. It is very important to identify such variant vessels and complex angioarchitecture before planning neuroendovascular or surgical intervention to prevent possible risks.

The relationships among Chiari malformation and persistent primitive arteries are not well understood. To date, there have been only three reported cases of primitive persistent arteries associated with Chiari malformation.\textsuperscript{[9,14,15]} The frequency of PPHA in Chiari malformation is also unknown. To the best of our knowledge, this is the first reported case of Chiari malformation in conjunction with PPHA and aneurysms. It remains unclear whether each lesion occurred independently or in association on the basis of altered/disturbed embryonal vasculogenesis. This case demonstrates that these pathologies can co-exist, highlighting the need to carefully analyze vascular architecture, focusing on each lesion.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References