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Case Report

Bilateral persistent primitive hypoglossal artery presenting with hemiplegia[☆]

Gagandeep Choudhary^a, Kriti Ahuja^{a,*}, Rihan Khan^b, Wayne Kubal^b^a Department of Radiology, University of Alabama in Birmingham, 619 19th Street South, JT 779, Birmingham, AL 35249-6835, USA^b Department of Radiology, Banner University Medical Center, Tucson, AZ, USA

ARTICLE INFO

Article history:

Received 28 February 2018

Revised 14 April 2018

Accepted 16 April 2018

Available online 22 May 2018

Keywords:

Persistent hypoglossal artery

Computed tomography

ABSTRACT

The manuscript describes an unusual vascular anomaly. Persistence of carotid-vertebrobasilar anastomosis is a rare occurrence with presence of bilateral hypoglossal arteries (HAs) rarer still. We present a case of bilateral persistent HAs with hypoplastic vertebral arteries which end into posterior inferior cerebellar arteries. The computed tomography and magnetic resonance imaging appearance, course, and other associations are discussed. A review of 6 cases of bilateral HA published in the world literature is also discussed.

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1. Case report

A 20-year-old female presented with sudden onset of right-sided headache and visual disturbance in the right eye which resolved within 2 hours and was followed by right arm and leg weakness with sensory deficit, right face numbness, and left-sided sharp chest pain. Clinical examination revealed mild weakness (4/5) and mild sensory deficit on the affected side with National Institutes of Health stroke scale of 3. The patient was hemodynamically stable with normal vitals. Stroke code was called and stat non-contrast computed tomography (CT) scan of the head and CT angiogram of neck and head vessels were obtained which revealed no hemorrhage or cortical infarction. The CT angiogram showed no evidence of vascular occlusion or stenosis. Bilateral hypoplastic vertebral arteries

directly ending into posterior inferior cerebellar arteries were seen. Anomalous vessels arising from the distal cervical internal carotid arteries (ICAs) at the level of C2 vertebral body and entering the skull through the hypoglossal canal were seen supplying the normal basilar artery (Fig. 1). She was further evaluated with magnetic resonance imaging stroke protocol which demonstrated no evidence of an infarct. Magnetic resonance angiogram also revealed anomalous vessels arising from the distal cervical internal carotid arteries and supplying the basilar artery with hypoplastic vertebral arteries (Fig. 2). Anterior inferior cerebellar artery and superior cerebellar artery were seen arising normally from the basilar artery. Bilateral posterior communicating arteries (PCOMs) were present. On further workup no cause of patient symptoms was identified. With time the patient's symptoms gradually improved and ultimately resolved. Given the normal workup with negative imaging findings and complete resolution

[☆] Conflict of Interest: None.

* Corresponding author.

E-mail address: kriahuja@uab.edu (K. Ahuja).<https://doi.org/10.1016/j.radcr.2018.04.022>1930-0433/© 2018 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license. (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

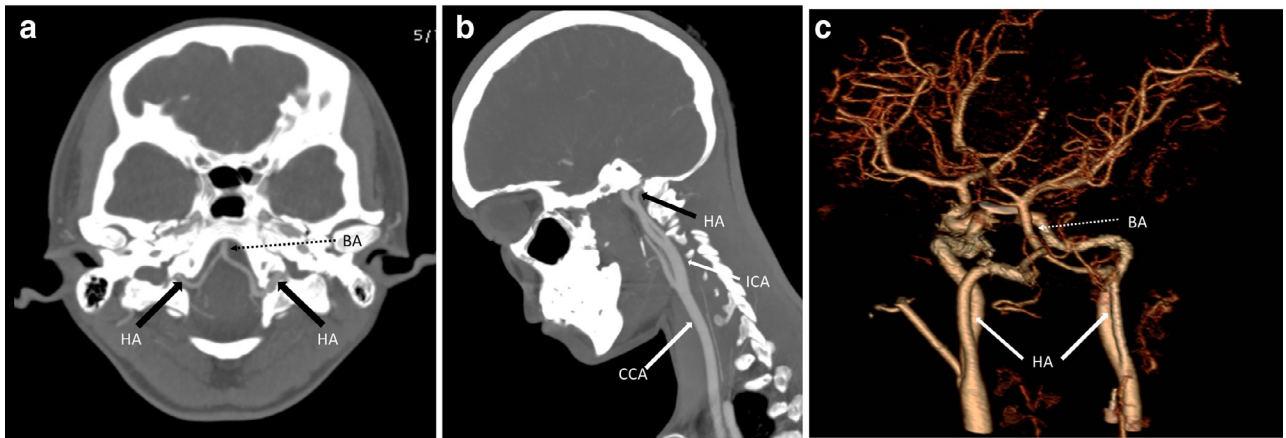


Fig. 1 – Axial maximum intensity projection (a), parasagittal maximum intensity projection (b) and three-dimensional rotational computed tomography angiogram (c) images for the brain and neck vessels. Persistent bilateral hypoglossal arteries (HAs) entering the skull through hypoglossal canals; basilar artery (BA) is formed by joining both HAs; common carotid artery (CCA) bifurcates to form internal carotid (ICA) and external carotid arteries; the internal carotid artery gives anomalous HA in mid cervical region.

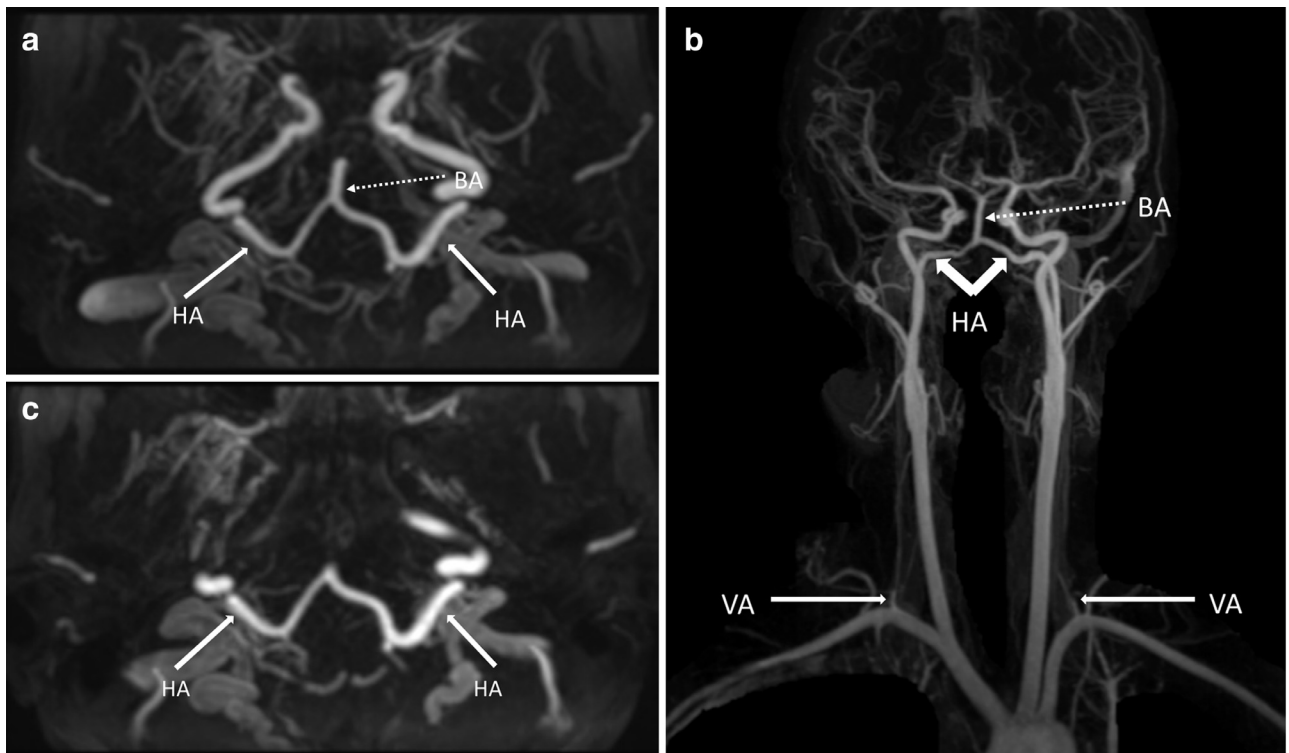


Fig. 2 – Axial magnetic resonance angiogram maximum intensity projection of intracranial arteries (a, c), coronal maximum intensity projection intracranial and cervical arteries (b). Persistent bilateral hypoglossal arteries (HA) entering the skull through hypoglossal canal; basilar artery (BA) is formed by joining both HA; bilateral hypoplastic vertebral arteries (VA).

Table 1 – Previously reported cases in the literature of bilateral persistent hypoglossal arteries.

Serial no.	Author	Age/sex	Vertebral arteries	Complication
Case 1	Murayama et al. [5]	59/M	Hypoplastic	Ruptured aneurysm located at the junction of left hypoglossal artery and basilar artery
Case 2	Takahashi et al. [2]	76/F	Absent	None
Case 3	Karasawa et al. [6]	39/M	Absent on left side Hypoplastic on right	Subarachnoid hemorrhage (cause was unknown even after autopsy)
Case 4	Oonishi [7]	-	-	-
Case 5	Garge et al. [8]	60/F	Both mildly hypoplastic	Subarachnoid hemorrhage with fusiform aneurysmal dilation of the left hypoglossal arteries toward its basilar segment
Case 6	Patira et al. [9]	79/M	Absent	Subocclusive thrombus in right cervical internal carotid arteries just distal to the hypoglossal arteries with infarct in right Middle Cerebral Artery (MCA) distribution
Case 7	(Our case)	20/F	Bilateral hypoplastic	None

of symptoms, the diagnosis of hemiplegic migraine was made and patient was discharged without any prescription with advice to follow-up with neurology.

2. Discussion

In the early human embryo, the forebrain is formed first which in turn establishes the anterior circulation obtained from the ICAs in the form of three presegmental arteries (trigeminal, otic, and hypoglossal arteries [HAs]), one permanent presegmental artery (PCOM) and the first intersegmental artery (also called proatlantal artery). These three presegmental arteries are named for their association with the trigeminal (fifth), vestibulocochlear (eighth), and hypoglossal (twelfth) nerves, respectively [1]. Few authors have also postulated that the PCOM is the fourth presegmental artery. All these arteries can be seen in embryo of 4-5 mm length. As the vertebrobasilar system and PCOMs develop these presegmental arteries obliterate, otic being first to obliterate followed by hypoglossal, trigeminal, and proatlantal artery successively [2].

Failure of obliteration of these arteries results in persistent of embryonic vessels, with overall incidence of 0.1%-1% [3]. The persistent trigeminal artery is most common followed by persistent HA (0.03% and 0.26%) [4], proatlantal artery and persistent otic artery being least common. These are frequently associated with the hypoplasia of the vertebrobasilar system. The presence of bilateral persistent HAs is very rare with just 6 cases reported in the literature till date [2,5–9]. Relevant findings of these cases from the literature and our case are described in Table 1.

Primitive HA follows the course of the hypoglossal nerve and connects the ICA to the primitive longitudinal neural artery which is a precursor of basilar artery [10]. The persistent HA arises from the ICA at C1-C3 level and enters the skull through the hypoglossal canal as in our case or rarely from anterior condylar canal to join with basilar artery. In cases with persistent HA the vertebral artery is absent or hypoplastic and

Table 2 – Criteria for identification of persistent hypoglossal artery (HA) [11].

1	HA must arise from the cervical part of the internal carotid artery at the C1-C2 level
2	Together, with the hypoglossal nerve, it must enter the posterior cranial fossa via the hypoglossal canal
3	Basilar artery must arise from a branch of HA
4	Ipsilateral vertebral artery may be hypoplastic

frequently ends in the posterior inferior cerebellar artery, as in our case. Criteria for identification of HA were postulated by Vlychou et al. [11] (Table 2).

Most of the times they are noted incidentally on scans done for unrelated indications without any direct clinical significance. Our patient's transient hemiplegia was not related to this anomalous vessel. The clinical diagnosis by the neurology team was hemiplegic migraine.

Rarely aneurysms can develop at the junction of basilar artery [5,8,12]. However, the incidence of vascular anomalies associated with the persistent trigeminal artery is up to 25% [4,13,14]. So, it is important to understand the anatomic variation with an increased incidence of other vascular malformations and potential risk associated with them.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at [doi:10.1016/j.radcr.2018.04.022](https://doi.org/10.1016/j.radcr.2018.04.022).

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