

Case Report

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Antonio Romeo*, Giuseppina Napolitano, Giuseppe Leone, Alessandra Aiello, Antonietta La porta, Enrico Tedeschi, Francesco Briganti, Ferdinando Caranci

A rare case of persistent hypoglossal artery associated with contralateral proximal subclavian stenosis

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Abstract: The persistent hypoglossal artery is rare vascular anomalies. We report the case of a 50-year old man with right hypoglossal artery, ipsilateral hypoplastic internal carotid artery, associated with left proximal subclavian stenosis with subclavian steal syndrome. Power-Doppler-Ultra-Sonography spectral images obtained after the patient exercised the left arm showed mid-systolic deceleration with retrograde late-systolic velocities. A Computed Tomography Angiography demonstrated a proximal stenosis of the left SA, a mild right ICA hypoplasia and an anomalous artery arising from right ICA at C2-C3 level, entering the cranium via the hypoglossal canal and joining the basilar artery. Usually the presence of PHA may be completely asymptomatic, and detected as an incidental finding by CTA or MRA, but in our case its diagnosis is extremely important because it is often the only vessel supplying blood to the basilar trunk and posterior circulation.

Keywords: Arteries, Abnormalities, Persistent primitive hypoglossal artery, CT angiography

*Corresponding author: Antonio Romeo, Department of Medicine and Health Sciences, University of Molise, Campobasso, via A. Mancini, 80127 Naples (Italy) E-mail: antromeo@gmail.com

Giuseppina Napolitano, Department of Medicine and Health Sciences, University of Molise, Campobasso, Italy

Giuseppe Leone, Alessandra Aiello, Enrico Tedeschi, Francesco Briganti, Ferdinando Caranci, Unit of Neuroradiology, Department of Advanced Biomedical Sciences, Federico II University of Naples, Italy
Antonietta La porta, Department of Radiology, Hospital Landolfi of Solofra, Avellino, Italy

1 Introduction

Persistent carotid-basilar artery anastomoses are rare vascular anomalies, including the trigeminal, otic, and hypoglossal arteries [1]. The persistent hypoglossal artery (PHA) is the second most common carotid-vertebrobasilar artery anastomosis after trigeminal artery, with a prevalence of 0.02%–0.10% [2]. We report on the case of a 50-year old man with right hypoglossal artery, ipsilateral hypoplastic internal carotid artery (ICA), associated with left proximal subclavian stenosis with subclavian steal syndrome (SSS).

2 Case report

A 50 year-old man was admitted to our Institution reporting dizziness, several episodes of syncope, dysarthria, severe memory problems and blood pressure differential between the arms to investigate a SSS.

The study was conducted at the Department of Radiology at the University Federico II of Naples, according to the principles of the Declaration of Helsinki and approved by the Ethics Committee of the University of Molise. Written informed consent was obtained from subject.

Color-Doppler-Ultra-Sonography (CD-US) spectral images which was obtained after the patient exercised the left arm (by opening and closing the hand for two minutes) showed mid-systolic deceleration with retrograde late-systolic velocities (Figure 1); this CD-US spectrum may resemble the profile image of a rabbit (the “bunny rabbit” sign) [3]. The left subclavian artery (SA) stole blood from the left VA to supply the ischemic arm; the right VA wasn’t demonstrated.

A Computed Tomography Angiography (CTA) with multiplanar (MPR) and Volume-Rendering (VR) reconstructions demonstrated a proximal stenosis of the left SA (Figure 2), a mild right ICA hypoplasia and an anomalous artery arising from right ICA at C2–C3 level, entering the

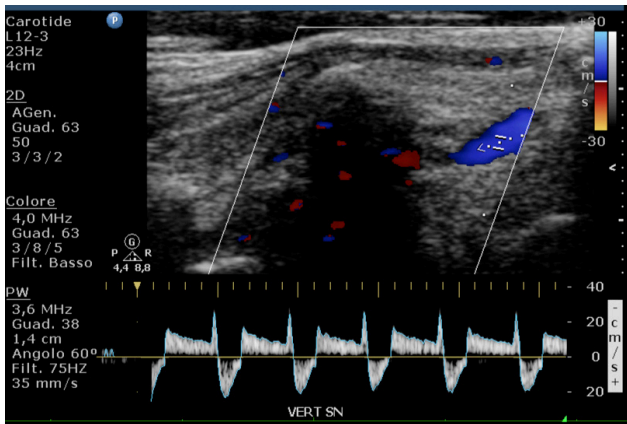


Figure 1: Color-Doppler-Ultra-Sonography spectral images obtained after the patient exercised the left arm (by opening and closing the hand for two minutes): mid-systolic deceleration with retrograde late-systolic velocities.



Figure 2: Computed Tomography Angiography (MPR reconstruction): proximal stenosis of the left subclavian artery (arrow).

cranium via the hypoglossal canal and joining the basilar artery (BA) (Figure 3, 4). The right hypoglossal canal was larger than the contralateral. There were no findings suggesting bony destruction of the hypoglossal canal.

The ipsilateral (right) vertebral artery was completely absent, while the contralateral left vertebral artery appeared regular. Due to the left SSS, the basilar artery was only refurnished by the right PHA.

3 Discussion

PHA originates from the internal carotid artery at the level of C1-C3 vertebral bodies, courses through the hypoglossal



Figure 3: Computed Tomography Angiography (MPR reconstruction): anomalous artery arising from the right internal carotid artery at C2–C3 level, entering the cranium via the hypoglossal canal.



Figure 4: Computed Tomography Angiography (VR reconstruction): the anomalous artery arising from the right internal carotid artery at C2–C3 level joins the basilar artery.

canal, and anastomoses with the basilar artery. Definitive diagnosis is based on the recognition of an anomalous artery in the enlarged hypoglossal canal [1,2].

According to Padget [4], during the early stage of embryological development the carotid system and two parallel longitudinal neural arteries supply the forebrain and the hindbrain, respectively. Four primitive anastomoses (named from the cranial nerves with which they course) occur between the carotid and vertebrobasilar systems: the trigeminal artery, the otic artery, the hypoglossal artery and the proatlantal artery. Normally, these anastomoses remain functional for approximately 7–10 days during the early stage of fetal development and then obliterate at the rate at which the posterior communicating arteries and vertebral arteries develop. Failure of this obliteration results in the persistence of embryonic

arteries, which leads to hypoplasia of the vertebrobasilar system [5].

Brismer [6] defined three essential diagnostic criteria in describing the PHA: the origin from ICA as an extracranial branch, the passage through the hypoglossal canal, the joining through the caudal part of the basilar artery. However, unusual and extremely rare variants of PHA have been reported, such as those arising from the external carotid artery (ECA) [7] or ending in the posterior inferior cerebellar artery (PICA) without an interposed segment of the basilar artery [8].

Uchino et al. [9] in a recent review proposed a new classification, naming “type 1” the usual PHA arising from the cervical ICA, “type 2” the PHA arising from the ECA, and “PHA variant” when the postero-inferior cerebellar artery (PICA) arises from the carotid system without connection to the VA.

The PHA is usually an incidental finding but its diagnosis is extremely important because it is often the only vessel supplying blood to the basilar trunk and posterior circulation; moreover, it could be associated to glossopharyngeal neuralgia [10], hypoglossal palsy [11], intracranial aneurysms [12]. In particular, a relationship between PHA and intracranial aneurysms has been reported, up to 26-33% in different series [9, 12].

In our case, all Brismer [6] diagnostic criteria were adhered to: the right VA was absent and the right ICA was hypoplastic. As demonstrated in large retrospective series [9], these findings suggest that the absence of the VA ipsilateral to PHA may result in the persistence of PHA. Left VA, as demonstrated by CD-US, showed retrograde flow due to proximal subclavian stenosis.

The left VA was functionally absent because of its retrograde flow and the vertebrobasilar system was only refurnished by the PHA. To the best of our knowledge, a similar anatomic and hemodynamic situation in a “type I” PHA (according to Uchino et al. classification [9]) has never been reported. Moreover, the anatomic and functional variant described in our patient is clinically important because both the anterior and posterior cerebral circulation are dependent on the arterial supply of the ICA, and the risk of bilateral occipital ischemia caused by embolism from the ICA to posterior cerebral arteries (PCA) through PHA is increased [13].

This suggests that it is important to remember that surgical and endovascular procedures in the ICA must be performed carefully in patients with PHA. This is because ICA cross-clamping during thrombo-endoarterectomy or its temporary occlusion during endovascular occlusion test [13-18] may significantly lower cerebral and brainstem perfusion.

4 Conclusion

The presence of PHA may be completely asymptomatic, and may be detected as an incidental finding by CTA or MRA. The recognition of PHA is essential to determine the therapeutic management of patients with atherosclerosis of the supraaortic vessels and vertebrobasilar ischemia.

Conflict of interest: Authors state no conflict of interest.

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