

An Unusual Cause of Syncope: Persistent Hypoglossal Artery, A Case Report

Senkop'un Nadir Bir Nedeni: Persistan Hipoglossal Arter, Olgu Bildirisi

ABSTRACT

Persistent hypoglossal artery is a rare primitive anastomose which mainly supplies the posterior cerebral circulation by itself in the case of the absent vertebral arteries. This artery is an embryological remnant and is commonly found incidentally. It originates from the upper cervical vertebra (C1-C2 level) of the internal carotid artery. Here we present a unique case suffering from syncope related to persistent hypoglossal artery which supplies the posterior cerebral blood flow in the absence of the vertebral arteries with the classical findings on magnetic resonance imaging and our clinical approach.

Keywords: Persistent hypoglossal artery, embryological variant, 3D, MR angiography

ÖZET

Persistan hipoglossal arter (PHA), vertebral arterlerin yokluğunda posterior serebral dolaşımı kendi başına besleyen nadir primitif bir anastomotik bağlantıdır. Bu arter bir embriyolojik kalıntı olup sıklıkla insidental olarak bulunmaktadır. İnternal karotis arter'in üst servikal vertebra (C1-C2) seviyesinden kaynaklanmaktadır. Bu vaka takdiminde vertebral arterleri olmayan ve posterior serebral kan akımını sağlayan PHA'yla ilişkili senkop gelişen bir olguyu manyetik rezonans görüntüleme'deki klasik bulgular ve klinik yaklaşımımız ile sunuyoruz.

Anahtar Kelimeler: Persistan hipoglossal arter, embriyolojik varyant, 3B, MR anjiyografi

Throughout embryologic life, fetal development is one of the astonishing processes of a human being, as well as the gradual pace and complexity of the growth. While these processes are ongoing, some remnants can be found incidentally, which can be the clue to the transition from primitive to advanced evolution. One of the rare examples is the persistent hypoglossal artery (PHA), which connects the carotid artery to the vertebrobasilar system. It has been evaluated as one of the multiple embryologic anastomoses. Although it is the second after the persistent trigeminal artery, the incidence is reported at approximately 0.1%, a rare variant of the posterior circulation system of the cerebrum.¹ Here, we would like to present a case regarding PHA as a reason for syncope with the characteristic imaging findings and clinical approach.


Case Report

A 25-year-old female was admitted to cardiology outpatient clinic. She had suffered from syncope, headache, and nausea, which were recurrent for 1 month. She complained that her attacks mainly occurred after she started the Pilates class, especially when she exercised in a semi-recumbent position. Her past medical history was unremarkable. She was alert, awake, and oriented to date, place, and person in her physical examination. Her complete neurological and cardiologic examination was regular. She had no nystagmus, nuchal rigidity, and cerebellar dysfunction. There was no pathology detected in her electrocardiography (ECG) (PR interval: 160 milliseconds, QRS interval: 90 milliseconds, heart rate (HR): 65/min, corrected QT interval (QTc): 416 milliseconds) (Figure 1A). According to ambulatory ECG, the mean heart rate was 72/min, and the mean QTc was 420 milliseconds. Also, there was no arrhythmia detected. Regarding repolarization abnormality and arrhythmic burden determination, T wave alternance (TWA) analysis was performed with a 24-hour ambulatory ECG using the modified

CASE REPORT

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Received: November 11, 2022

Accepted: November 29, 2022

Cite this article as: Mutlu D, Zambak Mutlu ÖP, Kültürsay B, Karagöz A. An unusual cause of syncope: Persistent hypoglossal artery, a case report. *Türk Kardiyol Dern Ars.* 2023;51(4):286-289.

DOI:10.5543/tkda.2022.8056



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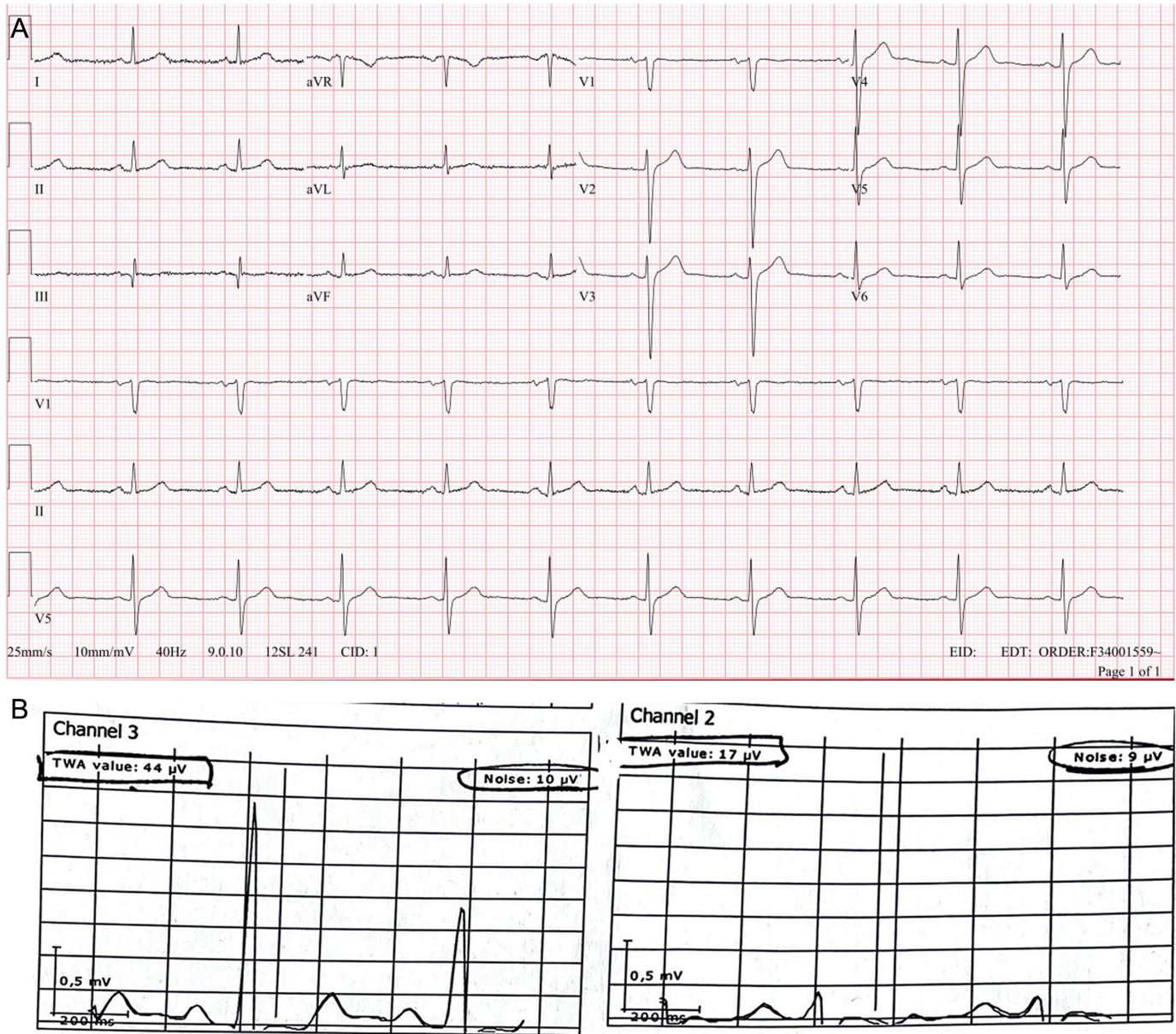


Figure 1. (A) The normal ECG recording, (B) TWA analysis with 24 hours ambulatory ECG. ECG, electrocardiography; TWA, T wave alternance.

moving average method (GE SEER 1000 ambulatory ECG and CardioDay analysis system, IL, Chicago, USA). The average TWA level was determined as $30.5 \mu\text{V}$, defined in normal ranges

(Figure 1B). Her transthoracic echocardiography (TTE) demonstrated that the left ventricular ejection fraction was 60%, and there were no valvular and structural abnormalities.

No significant changes in arterial saturation were found among positions and with exercise. Orthostatic hypotension and postural orthostatic tachycardia syndrome were also excluded by performing heart rhythm and blood pressure monitoring in lying down and standing-up positions. A colored Doppler carotid vertebral artery ultrasonography (USG) showed normal flow in carotid arteries, and bilateral vertebral arteries were not detected. Subsequently, 3-dimensional magnetic resonance angiography (MRA) and diffusion sequence were performed, revealing the absence of bilateral vertebral arteries. It was demonstrated that a vascular variation compatible with the PHA originated from the cervical segment of the right internal carotid

ABBREVIATIONS

BA	Basilar Artery
CT	Computed Tomography
ECG	Electrocardiography
PHA	Persistent Hypoglossal Artery
ICA	Internal Carotid Artery
MR	Magnetic Resonance
TTE	Transthoracic Echocardiography
USG	Ultrasonography
PCC	Posterior Cerebral Circulation
VA	Vertebral Artery

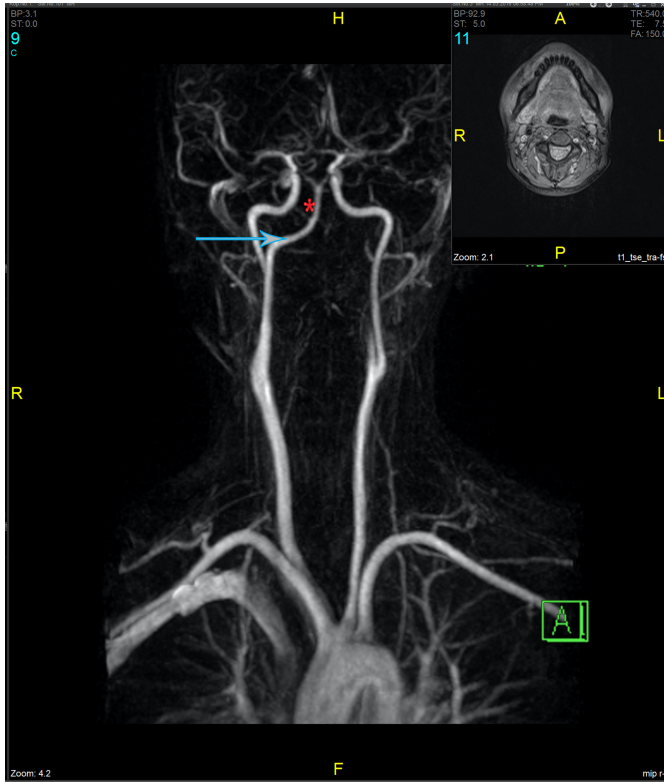


Figure 2. Three-dimensional reconstruction of the MRA. Arrow: PHA originates in the cervical portion of the C1-C2 level of the ICA. Asterisk: The BA is only filled by the anastomosis. Circle: Hypoplastic VA and PCA. MRA, magnetic resonance angiography; PHA, persistent hypoglossal artery; BA, basilar artery; VA, vertebral artery; PCA, posterior communicating artery.

artery (ICA) and provided blood flow to the basilar artery (BA). (Figure 2). Intracranial vasculature and bilateral common carotid artery were otherwise normal, and no intracranial pathology was revealed. All the results were evaluated in the multidisciplinary council with neurology and radiology as a benign condition, and it was decided to stop her exercise program and follow her yearly. At the 3-year follow-up, there was no complaint.

Discussion

Syncope is the rapid and short-duration unconscious state due to transient cerebral hypoperfusion and spontaneous recovery.² The etiology of syncope is not always easy to find. Although reflex-mediated syncope in young adults is the most common reason, cardiac or neurological pathologies and orthostatic hypotension can also cause syncope.² A multidisciplinary approach with systematic neurologic and cardiac evaluation is crucial to overcome this puzzling condition. When the first assessment is inconclusive, rare causes such as a transient ischemic attack, subarachnoid hemorrhage, or structural neurological diseases should be considered, and further assessment should be carried out.

In our patient, arrhythmic etiology was excluded by baseline, 24-h ambulatory, and orthostatic ECG recordings. In addition, TWA analysis was performed to reveal any repolarization abnormality or arrhythmic burden. There was no abnormality detected

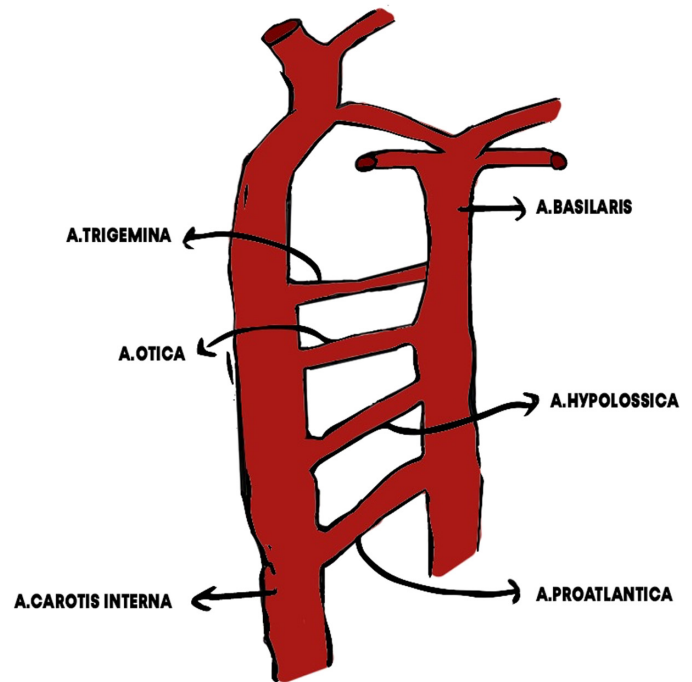


Figure 3. The figure illustrates the posterior primitive anastomoses. Arrow, persistent hypoglossal artery.

also the patient had no palpitations. Structural heart pathology was excluded by performing TTE.

Orthostatic hypotension and postural orthostatic tachycardia syndrome were also excluded by performing heart rhythm and blood pressure monitoring with lying down and standing-up positions, and also there was no significant QT interval change with orthostatic position according to the relevant guidelines for the management of syncope and ventricular tachycardia.^{2,3} Afterward, carotid and vertebral artery Doppler Ultrasonography (USG) was performed following the neurology consultation to investigate the underlying etiology and exclude internal causes. However, bilateral vertebral artery agenesis was demonstrated. Subsequently, MRA revealed PHA.

PHA, a rare primitive anastomose, mainly supplies the posterior cerebral circulation by itself in the case of the absent vertebral arteries. In embryologic development, when the caudal-cephalic length of the embryo is approximately 4 mm in the first week, the primitive dorsal aorta supplies blood to the fetal brain anteriorly and longitudinal neural arteries and vertebrobasilar anastomoses posteriorly.^{1,4,5} Posterior primitive anastomoses can be listed as, from proximal to distal, the trigeminal, otic, hypoglossal, and proatlantal arteries¹ (Figure 3). These are so-called because of their intimate association with the traces of cranial nerves or ganglions, such as the otic ganglion and hypoglossal nerve. These primitive arteries mainly supply the posterior cerebral blood flow in an embryo.

As the posterior circulation (PCC) system develops, these anastomoses become involute. The otic artery is the first to be obliterated, followed by the hypoglossal, trigeminal, and proatlantal arteries. If there is a regression problem, PCC has to be supplied by these primitive anastomoses according to their location.

This may intervene in developing aplastic vertebral arteries. It is generally originated from the upper cervical part of the ICA and connected to BA by using the hypoglossal canal.^{1,4} It is seen unilaterally in more than 90% of cases and slightly more common in women similar to our case.¹

Various diagnostic entities could be used for the diagnosis of PHA. Both computerized tomography and magnetic resonance imaging (MRI) angiography could reveal it triumphantly.⁴ Time of flight sequence of the MRI also could give valuable information without giving any contrast medium.⁶ The criteria of Brismar⁷ are used for the diagnosis in the evaluation process of angiograms. The criteria^{5,7} are as follows: (1) PHA originates in the cervical portion of the C1-C2 level of the ICA. (2) It passes through the hypoglossal canal to the posterior fossa. (3) The BA is only filled by the anastomosis. (4) Vertebral arteries and posterior communicating arteries are not visible by the radiographic techniques most of the time. In our case, the course of the artery was coherent with all of 4 criteria (Figure 2).

The migration of blood perfusion to the distal part of the body with adequate anterior flexion can be described as the link between PHA and syncope. As the gravity stimulation to the otoliths reduces the vasoconstrictive effect of neck muscle stimulation, Herault et al⁸ proposed that the vascular effect of otolith stimulation counteracts the neck-muscle stimulation in response to a fluid shift toward the legs.⁸ In the absence of manifest cerebral ischemia, Thierfelder et al. demonstrated relative hypoperfusion of the posterior cerebellar territory when assessed with delayed time to drain with CT, 42.4% of all screened patients with vertebral artery hypoplasia in the absence of manifest cerebral ischemia.⁹

Another possible explanation could be the Bow-Hunter syndrome, dizziness syndrome with flow interruption in the posterior cerebral circulation resulting from head movements.¹⁰ Cai et al¹⁰ reported a case with a right hypoplastic vertebral artery (VA) and a dynamically compressible left VA, managed surgically. Similarly, Choi et al¹¹ found that ischemia can be triggered by rotational head movements or tilting, leading to vertigo and syncope. As a result, excessive tilting of the head in the semi-recumbent position during dynamic pilates performance in PHA may have caused syncope, similar to exacerbation of ischemia.

To our knowledge, this is the second report in the literature regarding a patient experiencing syncope as a result of PHA. Jackson et al¹² reported a case of syncope directly related to this rare anatomic variant. In this report, syncope occurred due to extreme and sudden rotation of the head of the patient, comparable to Bow-Hunter syndrome. Our case differs from that because our patient did not experience any symptoms throughout the head rotation. Syncope occurred in a semi-recumbent position.

In conclusion, this rare condition emphasizes the importance of thinking that rare anatomic variants can also occur and lead to clinical reflection.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – D.M.; Design – D.M.; Supervision – D.M.; Materials – Ö.P.Z.M.; Data Collection and/or Processing – Ö.P.Z.M.; Literature Review – Ö.P.Z.M., B.K., A.K.; Writing – D.M.; Critical Review – Ö.P.Z.M., B.K., A.K.

Declaration of Interests: The authors declare that they have no competing interest.

Funding: The authors declare that this study had received no financial support.

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