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Case Study

# Two Cases of Rare Basilar Hypoplasia

Ivana Štula, MD, PhD[a*], Katarina Novak, MD, PhD[b], Krešimir Čaljkušić, MD[c] and Tonći Batinić, MD, PhD[a]

[a] *Clinical Department of Diagnostic and Interventional Radiology, Clinical Hospital Split, Split, Croatia*
[b] *Clinical Department of Internal Medicine, Clinical Hospital Split, Split, Croatia*
[c] *Clinical Department of Neurology, Clinical Hospital Split, Split, Croatia*

**ABSTRACT**

In this article, we report two cases of basilar artery hypoplasia; the first case was a primitive trigeminal artery, and the second was an isolated basilar artery hypoplasia. Both patients had general neurologic disturbances, including periodic intention tremor of the left hand. Our data underscore the utility of complementary time of flight (TOF) magnetic resonance and multidetector computed tomography (MDCT) angiography as reliable first detection methods for steno-occlusive diseases and in cases of suspected congenital vascular anomalies.

**RÉSUMÉ**

Ce texte présente deux cas d'hypoplasie artérielle basilaire; le premier est un cas d'hypoplasie primitive artérielle du trijumeau et le deuxième est un cas d'hypoplasie artérielle basilaire isolée. Les deux patients éprouvaient des difficultés neurologiques générales, dont un tremblement périodique involontaire de la main gauche. Nos données soulignent l'utilité de l'angiographie complémentaire TOF MR et MDCT comme méthodes de dépistage principales fiables des maladies occlusives et en cas de soupçons d'anomalies vasculaires congénitales.

*Keywords:* 3D TOF MR angiography; basilar artery hypoplasia; intentional tremor; MDCT angiography; persistent trigeminal artery

## Introduction

In early gestation, the paired ventral longitudinal neural arteries are connected to the internal carotid artery (ICA) by the posterior communicant artery (PCoA) and the primitive segmental arteries (trigeminal, proatlantal, hypoglossal, and otic). This vasculature constitutes the early prenatal posterior neural circulation. The primitive segmental arteries involute during the 5th week of gestation. In addition, the caudal end of the PCoA develops into two parts (P2 and P3) of the posterior cerebral artery (PCA), and the remaining PCoA shrinks. Finally, the basilar artery (BA) develops by fusion of the paired ventral longitudinal neural arteries on the anterior surface of the neural tube.

If the PCoA remains large, a PCA of "fetal origin" is said to persist. Aplasia or hypoplasia of the BA is the result of either an anomaly in the involution of primitive segmental arteries or fusion of the paired ventral longitudinal neural arteries. In a case of aplasia or hypoplasia of the BA, posterior circulation is supported by one of the primitive segmental arteries [1]. The most frequent persistent primitive anastomosis between the carotid and vertebrobasilar systems is the persistent trigeminal artery (PTA). This has been observed in 0.1%–0.2% of cerebral angiograms, usually as an incidental finding [2]. Segmental BA hypoplasia without persistent primitive segmental artery is extremely rare. Only eight cases were reported in literature [3, 4]. In this case, circulation of the PCA is mediated by the "fetal origin" PCoA.

### *Case 1*

A 67-year-old woman experienced general neurologic disturbances over the previous 5 years. Her first symptom was a slight tremor of the left hand. In the 2 years before examination, she also experienced episodes of transient global amnesia and, in the last few months, periodic ptosis of the right eyelid. Neurologic examination revealed only intention tremor of the left hand. No significant atherosclerotic plaques of the ICA were found nor was applanation of spectral waves observed on Doppler ultrasounds of the carotid and vertebral arteries. Psychological examinations showed cognitive deterioration of vascular etiology. Ischemic lesions in both frontal lobes

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* Corresponding author: Ivana Štula, MD, PhD, University Hospital Split, Spinčićeva 1, 21000 Split, Croatia.
E-mail address: ivanast23@yahoo.com (I. Štula).

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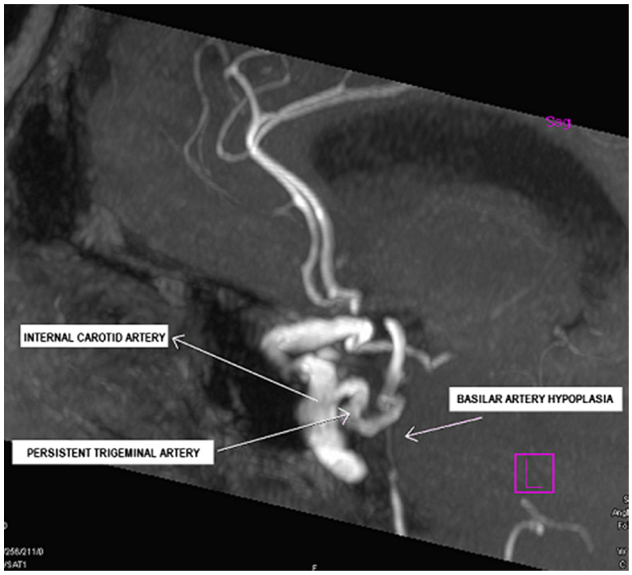


Figure 1. The hypoplastic distal part of the basal artery with persistent trigeminal artery.

and in the anterior aspect of the right capsula interna were detected by brain computed tomography (CT).

The patient was referred for three-dimensional (3D) time of flight (TOF) magnetic resonance angiography (MRA) of the brain (Avanto 1.5 T; Siemens, Erlangen, Germany; repitition time = 25 milliseconds, echo time = 7 milliseconds, flip angle = 25°, field of view = 180, matrix = 241 × 256, slice 120, slice thickness = 0.7 mm, slab 3, excitation 1) that revealed a hypoplastic distal 1.5 cm of the BA and communication with the right ICA through persistent PTA (Figure 1).

The persistent PTA supplied both superior cerebral arteries and the left PCA. The right PCoA supplied the right PCA.

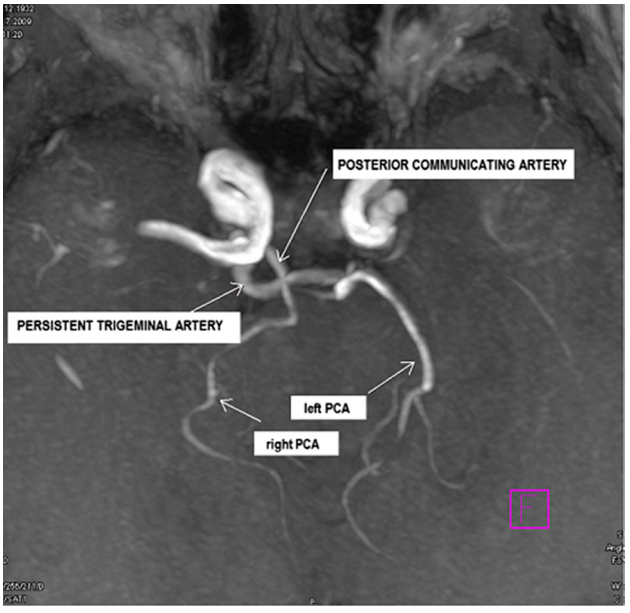


Figure 2. MRA of Saltzman type II of the trigeminal artery. PCA, posterior cerebral artery.

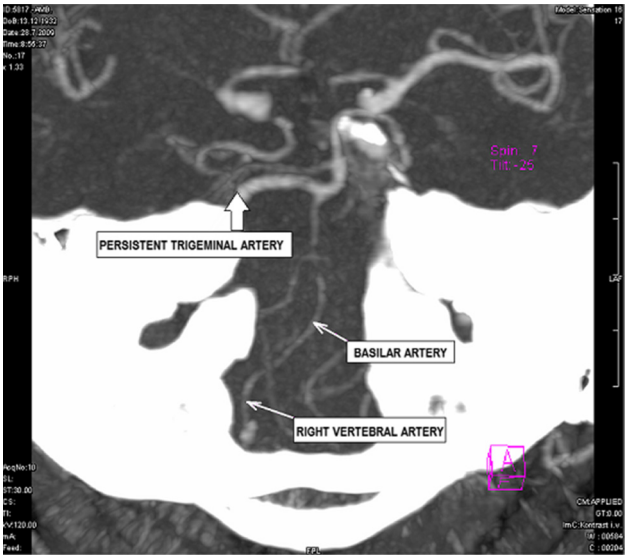


Figure 3. Hypoplastic BA with occlusion of the left VA.

Before that, the right PCA had an aplastic P1 segment (Saltzman type II of the trigeminal artery) (Figure 2).

The brain multidetector computed tomography (MDCT) angiography was performed using Sensation 16 (Siemens, Erlangen, Germany with the following acquisition parameters: slice of 1 mm, 120 kV, 100 mA, scan time of 4 seconds, and pitch of 1:15). The scan delay was set by an automatic bolus tracking technique after administrating an 80-mL bolus injection of nonionic contrast medium at a concentration of 350 mg/mL at the rate of 5 mL/s.

MDCT angiography showed hypoplasia of the entire BA with occlusion of the left vertebral artery (VA) and a hypoplastic right vertebral artery (Figure 3). MRA and MDCT revealed the same collateral circulation through the PTA and the right PCoA (Figures 2 and 4). MDCT did not reveal significant hyperdense plaque in the cavernous part of two ICAs, but it revealed absence of part of the sphenoidal sinus roof on contact surface with the right ICA (Figure 5a and b).

*Case 2*

A 74-year-old patient had been treated with phenobarbital for the last 15 years to control grand mal seizures. He had fractured his skull 5 years before examination. During the last month before admission, he experienced tremor of the left hand and one episode of speech disorder. An ambulant brain CT revealed hyperdensity in the left pontocerebellar angle, suggesting a VA malformation, but the MDCT was negative. The patient had an arrhythmic electroencephalogram with focal change on the left side. Neurologic examination revealed increased muscular tonus, intention tremor of the left hand, instability in the Romberg test, inarticulate speech, and bilateral hypoacusis.

After MDCT, the patient was referred for 3D TOF MRA, which was performed in the same manner as in the first case. A hypoplastic distal BA artery, beyond the branch point of the superior anterior cerebral artery, was revealed on 3D TOF