Bilateral Persistent Trigeminal Arteries with Unilateral Trigeminal Artery to Cavernous Sinus Fistula

A Case Report

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Summary

A 59-year-old man who denied a history of trauma presented with left pulsatile tinnitus and left orbital swelling for six months. Digital subtraction angiography showed a left persistent trigeminal artery (PTA) with a trigeminal artery to cavernous sinus (trigeminal-cavernous sinus) fistula and a right PTA. Transarterial detachable coil embolization of the left trigeminal-cavernous sinus fistula was performed, and the symptoms subsided. There has been no report of bilateral PTAs with a spontaneous fistula connected from one PTA to the ipsilateral cavernous sinus. This paper reports such a rare circumstance.

Introduction

One of the most common persisting embryological carotid-basilar anastomoses (CBAs), the incidence of unilateral persistent trigeminal artery (PTA) has been reported to be from 0.1% to 0.6% based on studies using large-scale magnetic resonance angiography (MRA) and digital subtraction angiography (DSA) imaging^{1,2}, and bilateral PTAs are relatively rare³⁻⁶. Trigeminal-cavernous sinus fistulas are uncommon lesions in which abnormal shunts between a PTA and an ipsilateral cavernous sinus develop spontaneously or secondary to trauma⁷⁻¹⁰. We describe a case of bilateral PTAs with a spontaneous trigeminal-cavernous fistula which we believe to be the first report of such a rare circumstance.

Case Report

A 59-year-old man with no history of trauma presented with left pulsatile tinnitus and left-orbital swelling for six months. He had been previously diagnosed to have a left indirect carotid artery to cavernous sinus fistula, and had received gamma-knife therapy at another hospital. However, the treatment failed to relieve his symptoms.

Six months later, the patient presented to our hospital. DSA imaging demonstrated that a right PTA originated from the cavernous segment of the right internal carotid artery (ICA), forming an anastomosis with the mid-basilar artery (Figure 1). Bilateral fetal-type posterior-cerebral arteries (PCAs) were also noted (Figures 1 and 2), suggesting that the right PTA was Saltzman type 2¹¹. An anomalous arterial branch emerged between the lacerum and petrous segments of the left ICA, which coursed upward along the left ICA, and connected to the left cavernous sinus, forming a fistula (Figure 2).

The anomalous branch was considered a caudad left PTA based on its proximity to the medial side of the V1 branch of the left-trigeminal nerve in the follow-up MRA, which is typical for a lateral-type PTA (Figure 4)^{1,12}. The trigeminal-cavernous sinus fistula was embolized transarterially (Figure 3) using Guglielmi detachable coils (Boston Scientific, Natick, MA, USA), and the symptoms subsided. Follow-up DSA and MRA examinations showed no recurrence of the fistula (Figure 4).

Figure 1 Angiography of right internal carotid artery showed the right persistent trigeminal artery (PTA) and the right fetal-type posterior cerebral artery (PCA). The right PTA was Saltzman type 2 based on the presence of the bilateral fetal-type PCAs.

Figure 2 Angiography of left internal carotid artery showed a caudad left PTA and the trigeminal artery to cavernous sinus fistula. The left fetal-type PCA is also noted. The superimposition of the caudad PTA and the precavernous internal-carotid artery led to an initial incorrect impression of an indirect carotid-cavernous sinus fistula.

Figure 3 After transarterial embolization of the left PTA and the trigeminal artery to cavernous sinus fistula, both were obliterated.

Figure 4 Follow-up time-of-flight MR angiography showed that the coils (arrow) were in close proximity to the left trigeminal nerve (asterisk). No recurrence of the left trigeminal artery to cavernous sinus fistula was visible. The right trigeminal artery persisted.

Discussion

Early in fetal development, the fetal carotid-basilar anastomoses (CBAs) connect the developing primitive internal carotid arteries with the paired longitudinal neural arteries that later fuse to form the basilar artery¹². The CBAs, which include the primitive trigeminal, otic, hypoglossal, and proatlantal intersegmental arteries, usually regress with the development of the vertebral and posterior communicating arteries (PCOMs)¹². Occasionally, however, these vessels persist after birth.

First described in an autopsy case by Quain in 1844 and angiographically by Sutton in 1950^{13,14}, the PTA is the most cephalad and the most common persistent CBA¹⁵, with a reported incidence from 0.1% to 0.6% based on large-scale MRA and DSA studies¹². A PTA usually originates from the cavernous ICA (C4 segment), proximal to the point at which the ICA exits the carotid canal near the posterior genu¹, and typically course posteriorly to the basilar

artery by either a parasellar or intrasellar course¹⁶. A parasellar (lateral-type) PTA curves posterolaterally around the dorsum sellae along the trigeminal nerve, traversing the dura medial to the V1 branch of the trigeminal nerve (Figure 4)^{1,16}. An intrasellar (medial type) PTA passes posteriorly, traversing the dorsum sellae and connecting with the basilar artery at the midline¹⁶.

Reports also exist of PTAs originating from the precavernous ICA, approximately 1.5 cm below the carotid siphon¹⁷. Other authors have described PTAs originating from the supraclinoid internal carotid artery^{17,27}. The left PTA in our patient was particularly caudad because it originated from between the lacerum and petrous segments of the left ICA (Figure 2). An anomalous branch emerging from the petrous ICA may be a persistent otic artery. However, this was excluded in our patient because the arterial branch followed the trigeminal nerve, rather than passing through the internal auditory canal¹⁸⁻²⁰. Moreover, an anomalous cere-

Table 1 Clinical and angiographic features of bilateral persistent trigeminal arteries reported in the literature.

Authors, Year, (Reference)	Age (y), Sex	Symptom & sign	PTA type	Associated vascular anomaly
Binet et al., 1977 ³	60, F	Headache, unsteady gait, cerebellar tumor	Bilateral PTAs	None
Taguchi et al.*, 1978 ²²	45, F	Diplopia, exophthalmoses	NA	NA
Okada et al., 1992 ²³	43, M	Left hemiparesis, brainstem infarct	Right PTA; Left PTAV to left SCA	Bilateral absent VA at origin
Chen et al., 1993 ²⁴	64, F	Severe headache, IVH	NA	Moyamoya disease
Alleyne et al., 1997 ²⁵	57, F	Headache, neck stiffness, SAH	Right PTAV to a right AICA/PICA common trunk; Left PTA	Right ACOM aneurysm, left pericallosal artery aneurysm
Ali et al., 2007 ²⁶	55, F	Incidental	Bilateral PTAs	Bilateral ICA aneurysm
Meila et al., 2011 ⁴	24, F	Right CN III palsy	Right PTAV to right PICA; Left PTA	Right PCOM aneurysm
Son et al., 2011 ⁵	46, F	Trigeminal neuralgia	Right PTA; Left PTAV to a left AICA/PICA common trunk	Bilateral vertebrobasilar junction hypoplasia
Uchino, 2011 ⁶	81, F	Incidental	Right PTAV to right AICA; Left PTAV to left AICA	None

PTA, persistent trigeminal artery; PTAV, persistent trigeminal artery variant; SCA, Superior cerebellar artery; VA, vertebral artery; IVH, intraventricular hemorrhage; SAH, subarachnoid hemorrhage; AICA, anterior inferior cerebellar artery; ACOM, Anterior communicating artery; ICA, Internal carotid artery; CN III, third cranial nerve (oculomotor nerve); PICA, posterior inferior cerebellar artery; PCOM, Posterior communicating artery; NA, Not available; *Article in Japanese.

bellar artery originating from the intracranial ICA with no connections to the basilar artery is classified as a persistent trigeminal artery variant (PTAV)^{1,2}. Since the first PTAV was reported by Teal et al. in 1972²¹, numerous anatomical PTAVs have been described^{1,2}. The course of PTAVs can be similar to any of the three cerebellar arteries. Incidences of 0.18% and 0.76% have been reported for PTAVs based on MRA and DSA studies^{1,2}.

Bilateral PTAs are an extremely rare condition. The eight cases reported in the English language literature (Table 1)^{3-6,22-26} were associated with intracranial aneurysm^{4,25,26}, Moyamoya disease²⁴, bilateral vertebral arteries agenesis, or hypoplasia^{5,23}. Among these, four had a classic PTA with a contralateral PTAV. One case with bilateral PTAVs was also reported⁶. It is possible that the arterial branch supplying the left trigeminal-cavernous sinus fistula in our patient was a left PTAV that connected to the cerebellar artery. The diameter of the distal branch of the left PTA/PTAV likely decreased because of the reduction in blood flow caused by the fistula shunt and previous gamma-knife therapy. Therefore, the distal segment of the left PTA/PTAV beyond the level of the left trigeminal-cavernous sinus fistula was not opaque in the left ICA and bilateral vertebral artery angiograms of our patient.

The angiographic anatomy and classification of PTAs were first described by Saltzman et al.¹¹. In Saltzman type 1 PTAs, the basilar artery proximal to the insertion of the PTA may be hypoplastic, and the bilateral PCOMs may be absent^{11,28}. Thus, a Saltzman type 1 PTA may be the only feeding artery for the distal branches of the basilar artery. The insertion of Saltzman type 2 PTAs on the basilar artery is below the bifurcation of the superior cerebellar artery, and the PCA receives blood predominantly through the patent PCOMs^{11,28}, whereas the basilar artery completely fills from one or both vertebral arteries. Thus, the Saltzman type 2

PTA is not critical to the maintenance of the cerebellar blood supply. Ali et al. classified PTAVs as Saltzman type 3 anomalies, and grouped them according to the insertion of the PTAV as Saltzman type 3a (superior cerebellar artery), type 3b (anterior inferior cerebellar artery), or type 3c (posterior inferior cerebellar artery)²⁹.

Whether an increased risk of aneurysm or other pathologies is associated with PTAVs is unclear¹⁷. However, structural abnormalities may exist in the wall of a PTA that predisposes it to vascular rupture. We found no evidence of aneurysm in our patient, but the spontaneous development of a fistula between the left PTA and the ipsilateral cavernous sinus suggests the possibility of a previous aneurysm or spontaneous vascular rupture. The goal of treating a trigeminal-cavernous sinus fistula is to preserve vision, eliminate bruit, and avoid ischemic and hemorrhagic complications. Endovascular treatment has superseded open surgery to close the abnormal connection between the feeding artery and the cavernous sinus. Treatment is generally administered via the transarterial or transvenous route, using balloons, coils, and liquid adhesive agents^{7,9,10,30-31}. Treatment for Saltzman type 1 PTAs should spare the PTA to prevent ischemia of the posterior circulation, whereas Saltzman type 2 PTAs can be embolized²⁸. The left PTA and the trigeminal-cavernous sinus fistula were simultaneously embolized in our patient.

Conclusion

This is the first report of bilateral PTAs presented with a spontaneous trigeminal-cavernous sinus fistula. The left PTA and the morbid trigeminal-cavernous sinus fistula were embolized transarterially using detachable coils. There was no complication or recurrence of the fistula following the procedure.

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