Fusiform aneurysm of a persistent primitive trigeminal artery associated with cerebrovascular anatomic variations: a report of two cases

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Abstract

There are four embryonic anastomoses that exist between the internal carotid artery (ICA) and the vertebrobasilar (VB) system, which may fail to regress postfetal life; one of which is the trigeminal artery. Other persistent anastomoses include those formed by the hypoglossal, otic and proatlantic intersegmental arteries. In addition, other cerebrovascular variations may accompany a persistent primitive trigeminal artery (PPTA); such as arteriovenous malformations, aneurysms, carotid-cavernous fistulas and stenotic cerebral vessels. We present two very rare cases of a left PPTA. In the first case, there was a past medical history significant for cervico-thoraco-lumbar spondylitis and in the second case of an operated occipital astrocytoma. In both cases, the PPTA was associated with a fusiform aneurysm located in the carotidian (lateral) aspect of the PPTA as well as other cerebrovascular anatomic variations. In the first case, the length of the PPTA was 26.0 mm and its endoluminal diameter, at its origin at the ICA, was 1.8 mm; the aneurysm has a length of 8.4 mm. In the second case, the length of the PPTA was 31.0 mm and its endoluminal diameter at its origin at the ICA was 3.0 mm; the aneurysm have a length of 7.6 mm. Identification of these variant vessels should not be ignored before planning and execution of neurosurgeries to prevent possible perioperative risks.

Keywords: persistent primitive trigeminal artery, fusiform aneurysm, anatomy, variation, associated neurological pathology.

Introduction

Richard Quain, in 1844, was first to give an account of a persistent primitive trigeminal artery (PPTA) [1, 2], which is an anastomosis between the internal carotid (ICA) and verteobasilar (VB) system. Other persistent anastomoses include those formed by the hypoglossal, otic and proatlantic intersegmental arteries [3–6]. A PPTA is the most common of the carotid–VB primitive anastomoses representing 80–85% of those reported and has an incidence varying between 0.06% to 0.76% [3, 5]. In the embryo, the trigeminal artery joins the cavernous segment of the left ICA with the distal parts of the basilar artery (BA) [2] and is found between the anterior inferior cerebellar (AICA) and superior cerebellar arteries (SCA) [1]. Tubbs et al. [1] found that 50–59% of PPTAs course through the sella turcica close to the clivus before connecting to the BA. Alternatively, the vessel can pass lateral to the sella turcica before joining the BA.

A PPTA usually presents incidentally on magnetic resonance imaging (MRI) angiography [7] and may be associated with other vascular abnormalities, such as abnormalities of the vessel’s wall, which can predispose to aneurysm formation [8]. An aneurysm of the trigeminal artery was first described by Davis et al., in 1956 [9] and an aneurysm of a PPTA that originated from the ICA was first reported by Bossi & Caffaratti in 1963 [10]. Wolpert [11] described a case of a PPTA aneurysm that was located at its attachment to the BA.

We report two cases of a PPTA identified incidentally via MRI angiography that was associated with a fusiform aneurysm of the carotid or lateral segment of the PPTA and other intracranial vessel variations.

Case reports

Case No. 1

We report a case of a 59-year-old male with a past medical history of cervico-thoraco-lumbar spondylitis who presented to the neurological emergency unit with a three-week history of sudden onset headaches and facial pain. A clinical examination was performed at the Neurosurgical Clinic of the Emergency County Hospital of Timișoara and completed with a MRI angiographic study at the Neuroimaged Diagnostic Imaging Centre, Timișoara (Romania), using a 1.5 T MR scanner. In addition to the spinal lesions, MRI angiography showed a left lateral PPTA arising from the cavernous segment of the left ICA and terminating at the
middle third of the BA. A fusiform aneurysm was located at the carotidian part of the PPTA.

The length of the PPTA was 26.0 mm and its endoluminal diameter, at its origin at the left ICA, was 1.8 mm. There was narrowing of the lumen of the PPTA for 1.2 mm after which there was a poststenotic dilatation, a fusiform aneurysm, which was 8.4 mm in length. The artery traveled another 17.6 mm with an endoluminal diameter of 2.0 mm, taking a medial course to supply the BA. The endoluminal diameter of the left ICA was 5.2 mm. The endoluminal diameter of the BA was 3.3 mm at its attachment to the PPTA. In addition, MRI angiography detailed the following: (1) a tortuous course of both vertebral arteries (VAs) a fusiform enlargement of the left VA and a hypoplastic right VA; (2) posterior cerebral arteries (PCAs) and AICAs that arose from the distal end of the BA; (3) a right PICA that originated from the fusiform enlargement of the left VA; and (5) agenesis of the right and left posterior communicating arteries (PCoAs) (Figure 1, A–C).

Case No. 2

We also report a case of a 31-year-old female with a past medical history of an occipital astrocytoma that was resected seven years ago. The occipital craniotomy left a right supra- and infratentorial porencephalic cavity, which involved the right occipital lobe and both cerebellar hemispheres equally. At three and six years post-surgery, she presented for routine follow-up without tumor recurrence. Clinical examination was performed at the Neurosurgical Clinic of the Emergency County Hospital of Timişoara and MRI angiography at the Neuromed Diagnostic Imaging Centre, Timişoara, using the 1.5 T MR scanner. Imaging revealed extensive temporo-occipital gliotic changes, focal cerebral atrophy and ectasia of the fourth ventricle. The MRI angiography also demonstrated a left lateral PPTA connecting the cavernous part of the left ICA to the distal third of the BA. The left PPTA had a length of 31.0 mm. At its origin, the PPTA had an endoluminal diameter of 3.0 mm and the fusiform aneurysm was located at the carotidian part of the vessel in “S” italic form with a length of 7.6 mm. The vessel maintained a medial course for 23.4 mm with an endoluminal diameter of 2.4 mm before connecting to the BA. The left ICA had an endoluminal diameter of 4.9 mm at the origin of the PPTA. The basilar artery had an endoluminal diameter of 3.6 mm at its medial connection to the PPTA. In addition, MRI angiography detailed the following: (1) an extremely tortuous course of both vertebral arteries (VAs) with a hypoplastic left VA; (2) left and right posterior cerebral arteries (PCAs) that arose from the distal end of BA (3) hypoplastic AICAs; (4) hypoplastic right and left PICAs that originated from the distal third of BA; and (5) agenesis of the left posterior communicating arteries (PCoAs) (Figure 1, D–F).

Discussion

Embryology

Padget [12] studied the primitive ICA at the 3 mm human embryonic stage where it starts as a cranial extension from the dorsal aortas. At the 5 to 8 mm stage, paired longitudinal neural arteries appear along the hindbrain and merge to form the basilar artery (BA). At the 5 to 6 mm stage, at ~29 days, the primitive PCoA forms and travels rostrally to connect to the primitive ICA and the paired longitudinal neural arteries [3]. In this period of development, the ICA supplies the forebrain, midbrain, and the hindbrain [2]. Prior to the development of the ICA, at the 3–5 mm stage, four pairs of fetal anastomotic arteries develop from the carotid and VB arteries at the level of the hindbrain. These anastomoses supply the posterior cranial circulation of the embryo and compensate for the absence/hypoplasia of the PCoAs or VAs [3, 6, 7]. These four primitive segmental anastomoses are, from caudal to cranial, the proatlantal intersegmental artery, hypoglossal artery, otic artery and trigeminal artery [13]. The trigeminal and otic artery branch off from the proximal segment of the ICA at the level of the trigeminal ganglion and otic vesicle respectively. The proatlantal and hypoglossal artery come from the paired dorsal aortas: the hypoglossal artery accompanied by the hypoglossal nerve and the proatlantal artery with the first cervical nerve [2].

The four pairs of fetal anastomotic arteries generally exist for seven to 10 days [3, 7, 14]. They subsequently regress by the 14 to 15 mm stage, at which time the PCoAs, BAs and the VB system is fully developed [3, 7]. According to Takahashi et al. [15], the otic artery regresses first, followed by the hypoglossal, the trigeminal artery and finally the proatlantal intersegmental artery. In rare cases, one of the primitive segmental anastomoses may persist into adult life. This variation is associated with absence or hypoplasia of the PCoAs and/or VAs [6, 7, 14]. The primitive trigeminal artery is the most common of the segmental anastomoses to persist, representing 83% of all persistent carotid–VB anastomoses. In extremely rare cases, the PPTA is also associated with a persistent otic artery [4], hypoglossal artery [16] or proatlantal intersegmental artery [17]. Bilateral PPTAs are extremely rare [18, 19].

Anatomy and classification

Typically, a PPTA is located on the posterolateral surface of the intracavernous ICA, nearer to the origin of the meningohypophyseal trunk [2]. It is much less common for a PPTA to arise more proximally from the petrous part of the ICA. Salzman [20] classified PPTAs according to the configuration of the vessel, the region the vessels supplies and its relationship to the PCoA. Type I variations describe PPTAs that supply the upper BA, PCAs, and SCAs. In this type, the proximal segment of the BA, at the insertion of the PPTA, may be hypoplastic or associated with an ipsilateral PCoA that is also hypoplastic or absent. Type II variations describe PCAs that receive their blood supply predominantly from a patent ipsilateral PCoA. In this configuration, the first segment of the ACA is absent and the PPTA becomes the main arterial supply to the territory of the SCA. Numerous variations based on the Saltzman’s classification exist. In Type III, according to Alcalá-Cerra et al. [2] and Son et al. [19], the PPTA does not combine with the BA but merges with the persistent longitudinal neural arteries to supply the cerebellar artery on the same side [2, 19]. Kim & Kim [21] describe five types based on the Saltzman’s classification.
Salas et al. [22] classified PPTAs into a medial (sphenoidal) or lateral (petrosal) type based on the vessel’s relationship to the abducens nerve. According to Uchino et al. [13], the lateral PPTA type originates from the cavernous segment of ICA near the posterior genu. It is surrounded laterally and posteriorly by the dorsum sellae and passes between the abducens (VI) and trigeminal (V) nerves to join the BA. The medial type of PPTA passes through the cavernous sinus and travels an intrasellar course usually in contact with the pituitary gland. Based on MRA examination, Weon et al. [23] classified PPTA into four types. In Type 1, the PPTA is the main blood supply to the distal BA, PCA and SCA territories (similar to Saltzman’s Type I). In Type 2, the PPTA supplies the SCA territories and the PCAs receive blood supply predominantly through a patent PCoA (similar to Saltzman’s Type II). In Type 3, the PPTA supplies the contralateral PCA. The ipsilateral PCA receives its blood supply through
the anterior circulation via the PCoA. In Type 4, the PPTA supplies the ipsilateral PCA and the contralateral PCA receives its blood supply though the anterior circulation via the PCoA. The cerebellar arteries that arise from the precentral segment of the ICA, without connecting to the BA, are described as PPTA variants [6].

**Incidence**

The PPTA is the most common of the primitive segmental anastomoses that connect the ICA and VB systems to persist into adult life, with a reported incidence between 0.06% [24] and 0.51% [13]. The persistent hypoglossal and proatlantal intersegmental arteries are the second and third most common variations with an incidence of 0.027–0.26% [6, 25] and 0.020% [1] respectively. The otic artery is the least common of the primitive segmental anastomoses to persist with an incidence of only 0.001% [26]. O’uchi & O’uchi [27], based on a study of 16 415 MR angiographic investigations, found 103 cases of PPTA and PPTA variants with an incidence of 0.34% and 0.29% respectively and a combined incidence 0.63%. In a study conducted by Uchino et al. [13], the cranial images obtained from 3491 MR angiographic studies revealed 12 cases of PPTA (incidence of 0.34%) and six cases of PPTA variants (incidence of 0.17%).

Uchino et al. [13] reported on 12 cases of PPTA that demonstrated a right/left ratio of 4:8 and a male/female ratio of 7/5. Kim & Kim [21] found a right/left ratio of 11:7 and male/female ratio of 5/13 in 18 cases of PPTA. Patient gender and right/left origin of the PPTA were not found to have any statistical significance. A review of the literature showed that bilateral PPTA variants are extremely rare (incidence: ~0.0012%) [18]. Early reports [5, 13, 21, 28] showed that the medial type of PPTA is also extremely rare. Uchino et al. [13] studied 3491 patients and found a medial to lateral ratio of 1:11, while Kim & Kim [21] in a study of 8900 patients found a medial to lateral ratio of 1:17. Kim & Kim [21] classified 18 cases of PPTA using the Weon et al. [23] classification and reported nine (50%) cases of Type 1, two (11.1%) cases of Type 2, six (33.3%) cases of Type 3, and one (5.6%) Type 4 case.

**Associations and clinical implications**

The PPTA and its variants may exist with other intracranial vascular anomalies or vascular diseases. The most common associated conditions are: tortuous bilateral internal carotid arteries [14], aplasia of the cervical and petrosal portions of the ICA [29], unilateral hypoplastic or aplastic VA [14, 27], a tortuous or dilated VA [14], termination of a VA into the ipsilateral PICA [27], hypoplasia of the basilar artery [3, 21, 28], the fetal type of PCoA [3], bilateral absence of the PCoAs [7], double middle cerebral artery [27], arteriovenous malformations [5, 7], carotid artery and cavernous sinus fistulas [5, 7], Sturge–Weber syndrome [5, 28], Moyamoya disease [28], vertebrobasilar embolic ischemia [28], kinking of the ICA [17], bilateral PPTAs [19], bilateral PPTA variants [18], persistent otic artery [4], ipsilateral Type 2 proatlantal intersegmental artery [30], bilateral proatlantal arteries [17], vascular nerve compression syndromes (such as trigeminal neuralgia) [19, 28] and cerebral aneurysms [4, 5, 7, 14, 31].

According to Tubbs et al. [1], PPTAs are associated with intracranial aneurysms in up to 14% of patients, but only 1% of these are aneurysms of the trigeminal artery. An early report by Cloft et al. [32] showed that the prevalence of intracranial aneurysms of the PPTA or a PPTA variant was 3–4.2%, which is similar to that of the general population. Aneurysms usually are saccular in form. Fusiform aneurysms are extremely rare [7, 31]. Most aneurysms occur in the medial type of PPTA, however they are extremely rare [5, 33]. The clinical implications of a PPTA aneurysm include cranial nerve palsies, carotid artery and cavernous sinus fistulas and subarachnoid hemorrhage, of these, subarachnoid hemorrhage is most common [5].

Our cases had PPTAs with associated fusiform aneurysms, tortuous course of both VAs, a hypoplastic VA (in one case the ipsilateral VA was affected and in the cases the contralateral VA was affected) and with absence of the posterior communicating arteries (PCoA) (in one case bilateral absence and in the other absence of only the contralateral PCoA). In both cases, the PPTA was an incident finding revealed during the management of cervico-thoraco-lumbar spondylitis and an operated occipital astrocytoma.

**Conclusions**

The PPTA is one of four primitive anastomoses that connect the internal carotid artery (ICA) to the vertebrobasilar (VB) system. In both of our cases, there was an associated fusiform aneurysm located in the carotid (lateral) part of the PPTA and other vascular pathologies. Identification of these variant vessels should not be ignored before planning and execution of neurosurgeries to prevent possible perioperative risks.

**Conflict of interests**

The authors declare that they have no conflict of interests.

**References**


O’uchi E, O’uchi T. Persistent primitive trigeminal arteries (PTA) and its variant (PTAV): analysis of 103 cases detected in 16,415 cases of MRA over 3 years. Neuroradiology, 2010, 52(12):1111–1119.


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