Persistent fetal intracranial arteries: a comprehensive review of anatomical and clinical significance

A review

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Intracranial fetal arteries that may be seen in the adult include, from anterior to posterior, the trigeminal, otic, hypoglossal, and proatlantal arteries (Figs. 1 and 2). These are named for their relationship with the trigeminal ganglion, otic vesicle, hypoglossal nerve, and dorsal aorta, respectively.12,57,69,70 When the human embryo is 4 mm long, these arteries serve as transitory anastomoses between the developing ICAs and bilateral longitudinal neural arterial plexus with the latter being a precursor of the future BA.12,48,49 Normally, these vessels are only of significance for approximately 1 week.21 The primitive trigeminal artery is the most commonly seen of these with an incidence between 0.1% and 0.5%. The primitive hypoglossal artery is the second most common with an incidence of about 0.1%, and the third most common is the proatlantal artery seen in 0.020%. The most uncommon anastomotic connection is the otic artery, seen in roughly 0.001%.5,42,89

Normally, once the PCoA forms, the presegmental arteries regress and become obliterated, beginning with the otic and followed by the hypoglossal, trigeminal, and proatlantal arteries.12 If these vessels persist, they may predispose to an increased risk of aneurysm73 or intracranial hemorrhage,58 although this topic is controversial. Additionally, these persistent carotid-vertebrobasilar anastomoses between the anterior and posterior cranial circulations are important to recognize during angiography for endovascular and surgical planning and should not be mistaken for other pathological conditions. Therefore, a thorough knowledge of their anatomy and potential clinical presentations is important to the head and neck surgeon and can help avoid unnecessary surgery and promote safe neurological surgery and interventional radiology.54,86 The aim of the present paper is to provide detailed descriptions of these vessels with a review of clinical and surgical implications.

Persistent Trigeminal Artery

The persistent trigeminal artery was first described by Richard Quain in 1844 (Figs. 1–4).70 In approximately 50%–59% of cases, persistent trigeminal arteries penetrate the sella turcica near the clivus to join the BA, and in the remaining cases the artery travels lateral to the sella turcica.35,71 The most common site of origin of the persistent trigeminal artery is the posterior bend or lateral wall of the intracavernous carotid artery.71 The connection to the carotid artery is usually just proximal

Abbreviations used in this paper: BA = basilar artery; ECA = external carotid artery; ICA = internal carotid artery; PCA = posterior cerebral artery; PCoA = posterior communicating artery; PICA = posterior inferior cerebellar artery; SCA = superior cerebellar artery; VA = vertebral artery.
to the cavernous sinus or just proximal to the meningohypophyseal trunk and to the BA between the SCA and anterior inferior cerebellar artery. The trigeminal artery may connect to the SCA or PICA and is then referred to as a persistent trigeminal artery variant. When present, the persistent trigeminal artery may be the main source of blood supply to the distal portion of the BA, SCA, and PCA. Importantly, the persistent trigeminal artery may give rise to up to 4 pontine perforating arteries.

The trigeminal and proatlantal arteries supply the cranial and caudal portions of the brainstem, respectively, in the embryonic stage, where they assume more important roles than the otic and hypoglossal arteries. By the 40-mm crown-rump length, the arterial supply of the head has approached its adult configuration. The persistent trigeminal artery may unite with the SCA or PICA.

Salas et al. described a case of a persistent trigeminal artery that pierced the dura mater between the abducent nerve medially and trigeminal nerve laterally. The authors distinguished 2 types of these fetal arteries regarding their relationships to the abducent nerve: a lateral or petrosal and a medial or sphenoidal. When the artery courses lateral to the abducent nerve intradurally, it arises from the posterolateral aspect of the intracavernous carotid artery. Passing inferior to this nerve within the cavernous sinus, the artery displaces the abducent nerve superiorly and then pierces the dura mater just medial to the sensory root of the trigeminal nerve. Conversely, when these arteries arise from the posteromedial aspect of the intracavernous segment of the carotid artery, they pierce the dura over the dorsum sellae and course medial to the abducent nerve, the so-called medial type.

Persistent trigeminal arteries have also been classified according to the configuration of the ipsilateral PCA. The Saltzman classification of persistent fetal trigeminal arteries includes 2 types; for the Type I variation, the BA proximal to the insertion of the trigeminal artery may be hypoplastic, and the PCoA may be hypoplastic or absent. Therefore, the trigeminal artery is the primary supply to the distal BA, PCA, and SCA territories. The Type II variation demonstrates a PCA that arises directly from the ICA and thereby allows the persistent trigeminal artery to fill the basivertebral system bilaterally (that is, the trigeminal artery joins the BA inferior to the origin of the SCAs, and the PCAs receive their blood supply primarily via a patent PCoA).

Numerous variations of the Saltzman classification exist. Suttner et al. reported a case of Saltzman Type I artery that originated from the intracavernous carotid artery and passed medial to the abducent nerve within a dural sleeve prior to joining the BA. Their case also demonstrated hypoplastic BAs and VAs distal to the junction with the persistent trigeminal artery. In cases of persistent trigeminal arteries, the VA and PCoAs are often hypoplastic ipsilaterally. This vessel may give rise to the meningohipophysial trunk. Tschabitscher and Perneczky stated that the persistent trigeminal artery is located superior to the oculomotor, trochlear, and abducent nerves and is medial to the ophthalmic part of the trigeminal nerve. Silver and Wilkins depicted the oculomotor, trochlear, and abducent nerves as coursing inferior to the persistent trigeminal artery. The third, fourth, and sixth cranial nerves have been compressed with resultant pare-
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Fig. 3. Angiogram of a persistent trigeminal artery.

sis in some patients with a persistent trigeminal artery. Some have stated that the persistent trigeminal artery arises from the ICA before it enters the cavernous sinus. This may not always be the case as we have illustrated with approximately one-half of the artery arising from the ICA traveling within the cavernous sinus. It has been found to branch from the petrous, or cavernous part of the ICA. A trigeminal arterial variant, originating from the cavernous ICA, can run along the fifth cranial nerve and enter the Meckel cave or through an isolated dural foramen, and can directly supply the cerebellum, without an interposing segment of the BA. Bhattacharya et al. described an anastomosis between the BA and ICA via trigeminal and stapedial artery remnants, as well as via the persistent otic artery.

Clinically, persistent trigeminal arteries may be associated with intracranial aneurysms in up to 14% of patients; however, only 1% of these are aneurysms of the trigeminal artery itself and whether these associations are simply incidental or not is controversial. Yang et al. reported a fusiform aneurysm of a persistent trigeminal artery variant. Komiyama et al. identified a persistent trigeminal artery in a patient with moyamoya disease. Trigeminal neuralgia has been found in patients with persistent trigeminal arteries. Reports also exist of a direct cavernous-carotid fistula involving persistent trigeminal arteries. Interestingly, Murtagh et al. reported a case of oculomotor nerve palsy found to be due to a persistent trigeminal artery. Ekinci et al. reported a case of hyperprolactinemia resulting from the compression of the pituitary stalk by a persistent trigeminal artery. Koch et al. postulated that a persistent trigeminal artery was responsible for compensation in a patient with subclavian steal syndrome.

Persistent Otic Artery

A persistent otic (acoustic) artery has been unequivocally documented at autopsy only once, and radiological observations are extremely rare with many of these reports being questioned by some authorities. In a review of 7382 carotid artery angiograms, the otic artery was observed only once. Lasjaunias and Berenstein stated that the otic artery cannot continue to exist without the persistence of the trigeminal artery, and that, unlike the other 3 carotid-vertebrobasilar anastomoses, there is no evidence for the existence of an otic artery in lower animals. Croft recently suggested that a true otic artery has yet to be reported. The otic artery may join the BA near the origin of the SCA or anterior inferior cerebellar artery or at the midportion of this vessel. The association with and potential derivation from the stapedial arterial system are debatable.
Pasco et al. stressed that the best identifying angiographic criterion for diagnosing an otic artery is to observe it coursing through the internal acoustic meatus. Hypoplasia or aplasia of the VA and PCoA may be seen in patients with an otic artery. Kaido et al. described fenestration of the ICA in the presence of an otic artery, and a trigeminal artery and a hypoglossal artery have been identified in patients with an otic artery. Symptomatically, a persistent otic artery has been reported as resulting in hemifacial spasm and paraparesis. We reported on a patient who suffered from left posterior circulation distribution infarctions following trauma. Angiography revealed a left ICA dissection. A reasonable conclusion for this case is that thrombus from the carotid artery dissection traveled via the otic artery to this patient’s posterior circulation.

As mentioned above, whether persistence of intracranial fetal vessels predisposes one to aneurysm formation is debatable, as the prevalence of such aneurysms is similar to the prevalence of aneurysms in the general population. Regarding the otic artery, Franz et al. described aneurysms of the left ICA and BA and an arteriovenous fistula in the presence of an otic artery. Simultaneous occurrence of an otic artery and aneurysms of the anterior communicating artery, PCoA, and cavernous segment of the ICA have been reported. In the case of Zhang et al., the otic artery arose from the cavernous ICA aneurysm and complicated endovascular therapy management.

**Persistent Hypoglossal Artery**

A persistent hypoglossal artery generally arises from the upper cervical portion of the ICA and traverses the hypoglossal canal to join the BA (Figs. 1C, 2, and 6–8). As the occipital somites are forming, the hypoglossal artery travels with the hypoglossal nerve from the cranium to neck, thus uniting the carotid and verteobasilar systems. In fact, an enlarged hypoglossal canal is important in making this diagnosis given that a persistent proatlantal artery takes a similar course but through the foramen magnum. This vessel was first described by Batujeff in 1889. Interestingly, Duffill et al. reported on a child who presented with subarachnoid hemorrhage from rupture of an aneurysm of a persistent hypoglossal artery. Yamamoto et al. reviewed the literature regarding aneurysms and persistent hypoglossal arteries and found that 26% of previously reported cases of this anomalous artery were associated with intracranial saccular aneurysms and most of these presented with rupture. This persistent vessel may be of functional significance in the patients with carotid artery stenosis and may provide a pathway for cerebral embolism.
Persistent Proatlantal Artery

The proatlantal artery emerges at the 4–5-mm embryonic stage and after the trigeminal artery regresses, serving as a main supplier to the posterior circulation of the brain together with the PCoA (Figs. 1D, 2, and 9). This vessel begins to regress at the 7–12-mm embryonic stage and is transformed into the horizontal segment of the VA as it courses over the posterior arch of C-1. It may arise from the upper cervical segments of the ECA or ICA. Some have also opined that the distal portion of the occipital artery may also be derived from this fetal vessel. In fact, some have found that this vessel has given rise to the occipital artery. Persistence of this vessel into adult life was first described by Gottschau in 1885. Proatlantal arteries have been classified into 2 types. Type 1 (proatlantal intersegmental) arteries arise from the caudal ICA and course between the arch of C-1 and the occiput to join the distal ipsilateral VA. Type 2 (C-1 intersegmental) arteries arise from the ECA and travel between the C1–2 interspace to join the ipsilateral VA. This type travels more laterally than the Type 1 artery and runs parallel to the horizontal portion of the VA. The origin of the PICA from the proatlantal artery has been reported, and the bilateral occurrence of the proatlantal artery in the adult is exceedingly rare.

Clinically, the importance of the anteroposterior view in differentiating the types of proatlantal arteries has been stressed by Obayashi and Furuse. The proatlantal artery has been associated with arteriovenous malformations. Although the proatlantal artery may be seen in conjunction with other aneurysms of the circle of Willis, aneurysms of this vessel have, to our knowledge, not been reported. Such persistent vessels in the upper cervical spine may predispose one to complications following surgical procedures in this area such as transarticular screw fixation. Additionally, care with endarterectomy or other procedures in the neck near the origin of these vessels should be considered.
Conclusions

Although uncommon, persistence of the intracranial fetal arteries may be encountered, the most common being the trigeminal artery. Although the literature is peppered with reports of the simultaneous occurrence of aneurysms in the presence of these vessels or involving them, no consensus has been reached as to whether this is an association or simply incidental. Clinically, ischemic presentations may be variable based on the presence of these vessels. Surgically, consideration of these vessels is important both for intracranial procedures as well as operations of the neck. Radiological examination of the vessels at the base of the skull and neck is important so as to avoid iatrogenic injury to persistent vessels.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Tubbs. Acquisition of data: Tubbs, Verma, Riech, Mortazavi. Analysis and interpretation of data: Tubbs, Mortazavi, Curé. Drafting the article: Tubbs, Verma, Mortazavi, Curé. Critically revising the article: Shoja, Loukas, Żurada, Cohen-Gadol. Reviewed final version of the manuscript and approved it for submission: all authors.

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Manuscript submitted September 3, 2010. Accepted November 30, 2010. Please include this information when citing this paper: published online January 14, 2011; DOI: 10.3171/2010.11.JNS101527. Address correspondence to: R. Shane Tubbs, M.S., P.A.-C., Ph.D., Section of Pediatric Neurosurgery, Children’s Hospital, 1600 7th Avenue South ACC 400, Birmingham, Alabama 35233. email: shane.tubbs@chsys.org.