



Case Report

A persistent trigeminal artery demonstrates cerebrovascular embryologic development

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ABSTRACT

Background: Cerebrovascular embryologic development is characterized by the presence of four well-described carotid-vertebrobasilar (VB) anastomoses. As the fetal hindbrain matures and the VB system develops, these connections involute, yet some may persist into adulthood. The persistent primitive trigeminal artery (PPTA) is the most common of these anastomoses. In this report, we describe a unique variant of the PPTA and a four-way division of the VB circulation.

Case Description: A female in her 70s presented with a Fisher Grade 4 subarachnoid hemorrhage. Catheter angiography revealed a fetal origin of the left posterior cerebral artery (PCA) giving rise to a left P2 aneurysm which was coiled. A PPTA arose from the left internal carotid artery and supplied the distal basilar artery (BA) including the superior cerebellar arteries bilaterally and the right but not left PCA. The mid-BA was atretic and the anterior inferior cerebellar artery-posterior inferior cerebellar artery complexes were fed solely from the right vertebral artery.

Conclusion: Our patient's cerebrovascular anatomy represents a unique variant of the PPTA not well described in the literature. This demonstrates how hemodynamic capture of the distal VB territory by a PPTA is sufficient to prevent fusion of the BA.

Keywords: Carotid-basilar anastomosis, Fetal intracranial artery, Persistent trigeminal artery, Trigeminal artery, Trigeminal

INTRODUCTION

The very first description of a persistent primitive trigeminal artery (PPTA) dates back to 1844 during an autopsy study by English anatomist and surgeon Richard Quain.^[8] A PPTA is the most common of the carotid-vertebrobasilar (VB) anastomoses: the incidence has been estimated to be 0.68% in a study examining magnetic resonance (MR) angiograms in over 15,000 patients.^[6] Numerous classification schemes have been developed to better characterize this anomaly. The Saltzman angiographic classification consists of two types.^[10] In the first, the PPTA inserts between the anterior inferior cerebellar artery (AICA) and superior cerebellar artery (SCA), supplying the distal VB circulation. The basilar artery (BA) proximal to the PPTA union is often hypoplastic and the posterior communicating arteries (PCoA) may be absent. In Type 2, the PTA supplies the SCAs and fetal-type posterior cerebral arteries (PCAs) supply the PCA

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territory. Unfortunately, this grouping is not all-inclusive and both a combination of Saltzman Type 1 and Type 2 as well as several Saltzman variants have since been discovered. This report describes a PPTA variant with a four-way division of the VB system. This variant has yet to be comprehensively documented in the literature and serves as an example of adult anatomy illustrating cerebrovascular development in the embryo.

CASE REPORT

History and examination

A female in her 70s with a history of hypertension and hyperlipidemia developed the sudden-onset of headache and slurred speech followed by unresponsiveness. Initial computed tomography (CT) imaging revealed a Fisher Grade 4 subarachnoid hemorrhage. There was prominent blood in the suprasellar, pre-pontine, and left perimesencephalic cisterns with an intraparenchymal component extending to the left thalamus, midbrain, and cerebellum. On transfer to our institution, her neurological examination was poor with evidence of a weak cough reflex and flexion of the lower extremities to painful stimulus.

Intervention and post-procedural course

After the placement of an external ventricular drain for the treatment of acute hydrocephalus, she underwent a diagnostic cerebral angiogram which revealed a multilobular left P2 aneurysm measuring 8.0 mm in maximum dimension [Figure 1]. There was a fetal origin of the left PCA. During catheter angiography for endovascular coiling, left internal carotid artery (ICA) angiogram showed a PPTA supplying the distal half of the BA, including the SCAs bilaterally and the right PCA but not the left PCA, which had a fetal origin. The proximal half of the BA up to the AICA-posterior inferior cerebellar artery (PICA) complex was isolated from the distal half by a mid-basilar atresia and was fed exclusively from the right vertebral artery (VA). The left VA was hypoplastic. Despite successful aneurysm coiling, the patient did not have significant clinical improvement. Given her persistent poor neurological examination over the next several days, the family decided to pursue comfort care measures. The patient ultimately expired on hospital day 2.

DISCUSSION

Embryologic origins

In 1948, Dorcas Hager Padget described the stages of cerebrovascular embryologic development in her publication titled “The development of the cranial arteries in the human embryo.”^[7] Much of our current understanding of neuroembryology comes from her tracings at the Carnegie

Institution of Washington in Baltimore depicting seven phases of neural arterial development in the embryo. At the 3 mm stage, the ICA and trigeminal artery branch from the first aortic arch. The trigeminal artery travels posteriorly to communicate with the vascular arterial plexus found on the ventral surface of the hindbrain which eventually forms the paired longitudinal arteries. At the 5–6 mm stage, the caudal ICA will anastomose with the rostral longitudinal neural artery forming the PCoA. Development of the definitive PCoA is typically accompanied by involution of the trigeminal artery. The means by which a primitive artery persists into adulthood has not yet been elucidated although it is thought that neurological development alters the hemodynamic balance leading to prominence of certain primitive branches, regression of others, and ultimately, the development of normal adult anatomy.^[5]

Classifications

As neuroimaging capabilities developed, multiple PPTA subtypes have been identified and characterized. The earliest angiographic classification was documented by Saltzman in 1959.^[10] In a study of eight cases, he described a first subtype with filling of the entire VB system distal to the anastomosis by the PPTA in conjunction with small or absent PCoAs. In the second subtype, the anastomosis primarily supplies the SCAs while the PCA territories receive blood supply from robust PCoAs. One patient manifested a combination of the two, whereby a fetal-type PCoA supplied the PCA territory on one side and the PPTA gave rise to the SCAs as well as the contralateral PCA. The term “Saltzman Type 3 PPTA” or “Saltzman variant” is given to a PPTA that terminates as a cerebellar artery without interposition of the BA.^[11] Type 3a, 3b, and 3c refer to direct termination as the SCA, AICA, and PICA, respectively. Although the Saltzman classification is frequently cited in the literature, some have questioned its relevance, suggesting that the supratentorial PCoA has a distinct embryologic origin from the infratentorial trigeminal artery.^[6,11]

Weon *et al.* proposed a more comprehensive classification based on MRA studies in a large cohort of patients.^[13] Type 1 and type 2 are analogous to Saltzman Type 1 and Type 2. A Type 3 corresponds to the aforementioned patient in Saltzman’s series: in this case, the PPTA supplies the contralateral PCA. In Type 4, the PPTA supplies the ipsilateral PCA and a designation of Type 5 is analogous to the three “Saltzman Variants” terminating directly as cerebellar arteries. Focusing solely on the anatomic course of the vessel as opposed to arterial relationships, Salas *et al.* classified the PPTA as lateral (petrosal) or medial (sphenoidal).^[9] In the lateral type, the artery arises from the posterolateral cavernous ICA, travels underneath the abducens nerve, and pierces the dura medial to the sensory

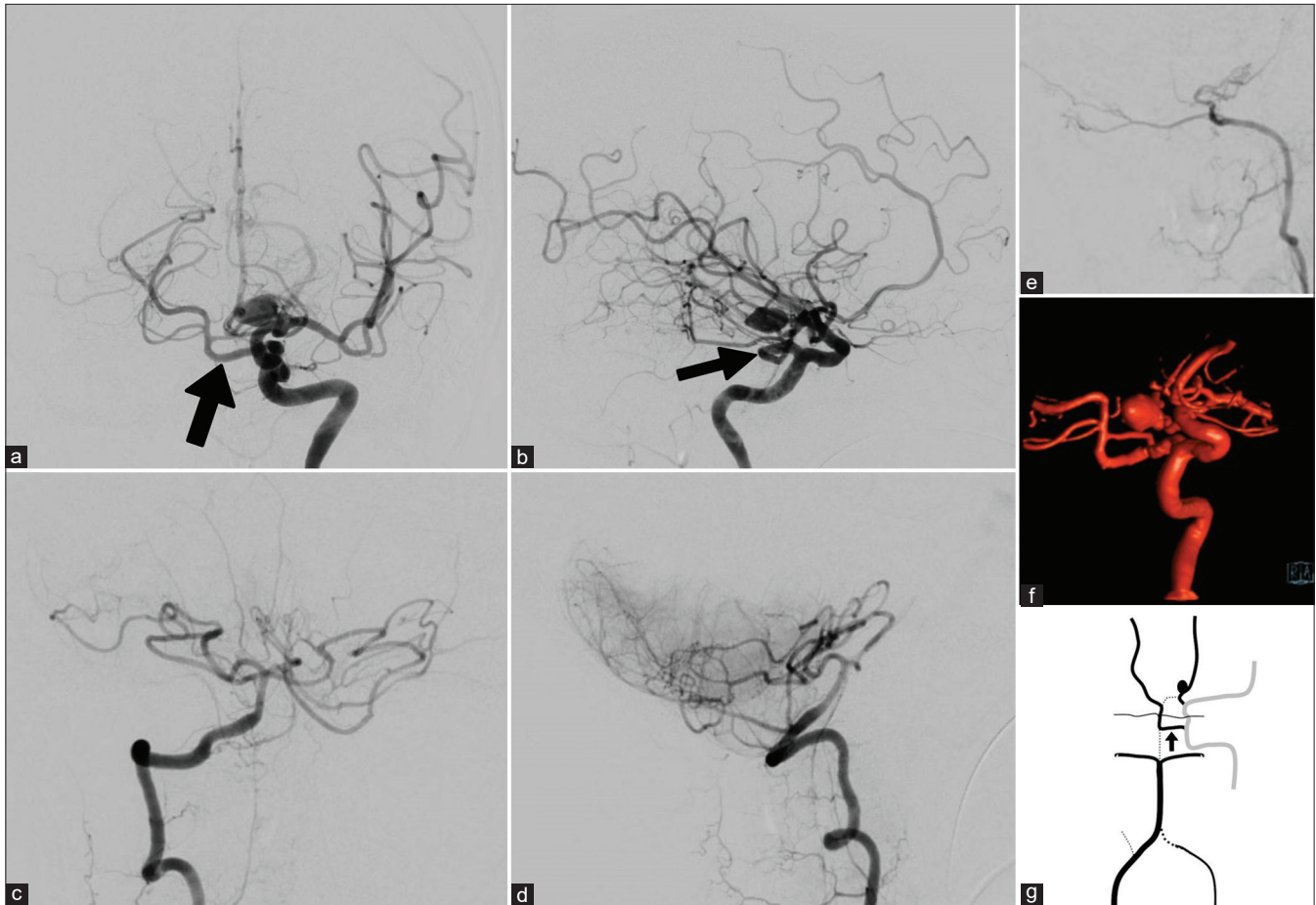


Figure 1: (a) Anterior-Posterior (AP) left internal carotid artery (ICA) angiography demonstrates a persistent primitive trigeminal artery (PPTA) (black arrow) supplying the right posterior cerebral artery (PCA) and SCAs bilaterally. (b) Lateral left ICA angiography further demonstrates the PPTA (Black arrow) and a left P2 aneurysm. (c) AP right vertebral artery (VA) angiography demonstrates the proximal basilar trunk with anterior inferior cerebellar artery - posterior inferior cerebellar artery complexes bilaterally. (d) Lateral right VA angiography further demonstrates the blood supply to the caudal cerebellum. (e) Lateral left VA angiography demonstrates termination after the sulcus arteriosus as meningeal branches. (f) Reconstructed oblique left ICA 3-dimensional rotational angiography demonstrates a left fetal-type PCA giving rise to the P2 aneurysm. (g) Diagrammatic representation of the patient's cerebrovascular system. Solid black lines represent blood vessels of the VB system while solid grey line represents the carotid circulation. Dotted black lines indicate a typical vessel that is absent. PPTA is indicated by black arrow.

root of the trigeminal nerve. In the medial form, the PPTA arises from the posteromedial cavernous ICA, passes medial to the abducens nerve and pierces the dura of the dorsum sellae. This may begin to relate vascular development with the concurrent neural and cartilaginous development, such as the interposition of the basal sphenoid cartilage.

Significance of basilar artery hypoplasia

In the case of vascular territory capture by a PPTA, less hemodynamic demand is placed on the BA proximal to the site of anastomosis. To this end, many PPTAs have been found in conjunction with basilar hypoplasia with a rate of approximately 42%.^[2,6] Basilar hypoplasia in of itself has been suggested to be an ancillary sign signifying the presence

of a PPTA.^[12] The spatial resolution provided by MR-angiography and CT-angiography often precludes a definitive differentiation of BA atresia from severe hypoplasia. Our case provides an angiographic demonstration of how hemodynamic capture of the distal VB territory by a PPTA is sufficient for incomplete fusion of the BA. This results in complete atresia which may not always be apparent in non-invasive imaging studies.

Association with cerebral aneurysms

A PPTA has been described in the context of several pathologies including cerebral aneurysms, carotid-cavernous fistulae, moyamoya disease, and trigeminal neuralgia among others. While early reports have cited a higher prevalence

of cerebral aneurysms in patients with a PPTA, more recent literature has suggested this finding to be the result of selection bias.^[3,4,6] Broadly speaking, cerebral aneurysm formation has been attributed to the compound effect of high wall shear stress at branch points of the circle of Willis and the structural deficiency of the tunica media at these locations. A tear in the internal elastic lamina at a bifurcation can more easily propagate, leading to aneurysm formation. As a PPTA inherently forms multiple branch points, it follows that opportunities for aneurysm formation may increase. However, at this time, there exists insufficient data quantifying changes in hemodynamics in the presence of a PPTA. Moreover, the relationship between a PPTA and a cerebral aneurysm in a different location, as in the present case, remains unclear.

Relevance to present case

Within the scope of current classification schemes, the present case most closely resembles a combination of a Saltzman type one and two. This particular variant is now more accurately described as a Weon Type 3 PPTA. The frequency of severe basilar hypoplasia commonly seen in this subset of PPTA is illustrative of the functional importance of the anterior circulation. A completely atretic BA, however, has yet to be well documented in the literature. Although configurations of the larger and more functionally important posterior circulation arteries have been well-studied, this is not the case with other branches of the VB system.

As the functional contribution of the vertebral arteries and proximal BA decreases, an array of anatomic variations in the posterior circulation may be seen.^[12] In these cases, hypoplastic VAs, VAs ending in PICA or PICA-AICA complexes are not uncommon. It is within this framework that our case importantly demonstrates a four-way separation of a typical VB system. The most common division of the VB territory occurs when an early fetal configuration persists of the PCA originating from the caudal carotid artery to supply the posterior circulation. A PPTA of any aforementioned classification represents a second division of the VB system. In our patient, a third division manifested as complete isolation of the AICA-PICA complexes bilaterally with no connection to the rostral BA. Finally, the left VA terminated after the sulcus arteriosus as meningeal branches.

CONCLUSION

The rarity of basilar segmentation is illustrative of the sequential stages of embryological development. As Hager Padget originally described, the regression of the primitive carotid-VB anastomoses occurs in an early embryonic stage

before the stochastic capture of vascular territories by the posterior circulation. Our case demonstrates how a PPTA can provide robust hemodynamic competition leading to incomplete fusion of the BA. The combination of a fetal origin of the PCA, a PPTA, isolated bilateral AICA-PICA complexes demonstrates a four-way division of a standard VB arterial network. We therefore find an example of adult anatomy representing the development of cranial arteries in the human embryo.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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