

Case Reports & Case Series

Endovascular treatment of a basilar tip aneurysm via a persistent primitive proatlantal intersegmental artery[☆]

Theresa A. Elder (MD)^{a,e,*}, Leonard H. Verhey (MD, PhD)^b, Devin B. Simon (MD)^c, Eleanor C. Smith (BS)^a, Joseph G. Adel (MD)^{a,d,e}

^a College of Medicine, Central Michigan University, 1280 East Campus Dr, Mt. Pleasant, MI 48858, USA

^b Division of Neurological Surgery, Department of Clinical Neuroscience, Spectrum Health, Michigan State University, 25 Michigan St NE Suite 6100, Grand Rapids, MI 49503, USA

^c Department of Neurology and Ophthalmology, Spectrum Health, Michigan State University, 2750 E Beltline Ave NE, Grand Rapids, MI 49525, USA

^d Ascension St. Mary's Hospital Neurosurgery Associates, 800 S Washington Ave, Saginaw, MI 48601, USA

^e Field Neuroscience Institute, 4677 Towne Center Rd #301, Saginaw, MI 48604, USA

ARTICLE INFO

Keywords:

Aneurysm
Basilar tip
Congenital
Proatlantal
Endovascular
Stent-assisted coiling

ABSTRACT

Background: A proatlantal intersegmental artery is an anastomosis between the carotid and vertebrobasilar arterial systems that exists in the developing fetus to temporarily maintain the posterior cerebral circulation. Failure of this artery to obliterate once the vertebral arteries are developed results in a persistent primitive proatlantal intersegmental artery (PPPIA). We present a patient with a basilar tip aneurysm that was exclusively supplied by a PPPIA, serving as the only corridor to accessing the aneurysm endovascularly. We report the first case of an elective endovascular stent-assisted coiling of a basilar tip aneurysm via a PPPIA and review this rare anatomic variant in a clinical setting.

Case report: The patient is a 60-year-old Caucasian male who presented with recurrent headaches and blurry vision after sustaining mild head trauma. Imaging revealed a basilar tip aneurysm, and the patient elected for endovascular treatment. He was furthermore found to have an anatomic variant of a proatlantal artery in the context of bilateral vertebral artery hypoplasia, thus his posterior circulation was entirely supplied by the fetal remnant vessel. The patient underwent a successful stent-assisted coil embolization of the aneurysm.

Conclusion: PPPIA is a rare vascular anomaly that may be associated with intracranial vascular pathology. Awareness and intricate understanding of the anatomy of embryologic remnants such as the PPPIA is central to planning neuro-interventional access and avoiding complications.

1. Background

The proatlantal intersegmental artery is one of four transient anastomotic vessels between the anterior and posterior cerebral circulation that forms at approximately thirty days gestation [1,2]. Together with the trigeminal, otic, and hypoglossal vessels, the proatlantal intersegmental artery maintains the posterior circulation via the primitive internal carotid arteries (ICA) until the vertebrobasilar system has matured at approximately seven weeks gestation [3]. The proatlantal intersegmental artery has been characterized into three types based upon its vessel of origin: Type 1 originates from the ICA, Type 2 emanates from the external carotid artery, and Type 3 is the rarest form

which originates in the common carotid artery [4]. Failure of the proatlantal intersegmental artery to involute during embryologic development results in a persistent primitive proatlantal intersegmental artery (PPPIA) [5]. Several reports of PPPIA have been published [6–12]. The co-occurrence of a PPPIA and an intracranial aneurysm is relatively high, and has been described in as many as 10% of the cases of PPPIA [13]. We report the first case of a patient with both a PPPIA and a basilar tip aneurysm treated with stent-assisted coil embolization via the unilateral PPPIA, it being the sole corridor to the vertebrobasilar circulation.

Abbreviations: CT, computed tomography; ICA, internal carotid artery; PPPIA, primitive persistent proatlantal intersegmental artery; PCA, posterior cerebral artery

[☆] This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

* Corresponding author at: 4677 Towne Center Rd, Suite 301, Saginaw, MI 48604, USA.

E-mail addresses: elder1ta@cmich.edu (T.A. Elder), leonard.verhey@spectrumhealth.org (L.H. Verhey), devin.simon@spectrumhealth.org (D.B. Simon), smith7ec@cmich.edu (E.C. Smith), joseph.adel@ascension.org (J.G. Adel).

<https://doi.org/10.1016/j.inat.2019.100543>

Received 17 June 2019; Accepted 21 July 2019

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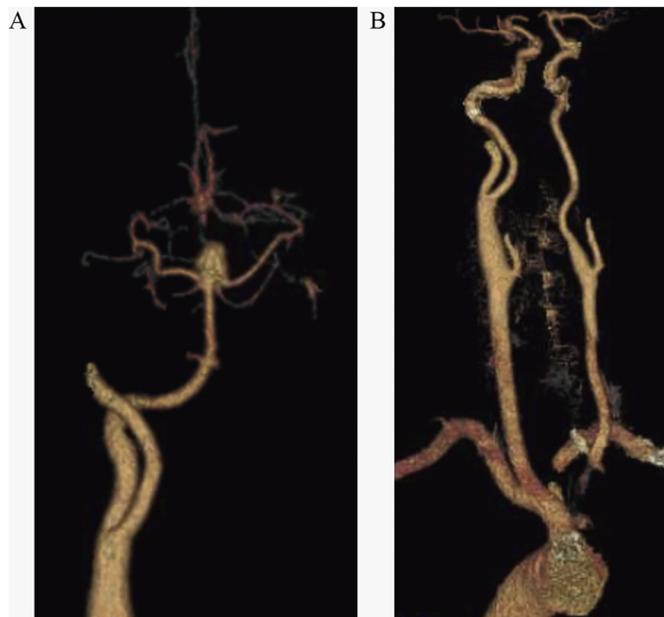


Fig. 1. CT Angiography, three-dimensional reconstruction. A) Right ICA reconstruction demonstrating the type one PPPIA and the basilar tip aneurysm. B) Bilateral carotid and subclavian reconstruction revealing the type one PPPIA and bilateral vertebral artery hypoplasia.

2. Case report

The patient is a 60-year-old Caucasian male, previous smoker, with family history of ruptured intracranial aneurysms, who sought medical attention at a local emergency department due to persistent headache after sustaining a minor head trauma a few weeks prior. Computed tomography (CT) of the head was suspicious for a basilar tip aneurysm, for which the patient was transferred to our facility for management.

On arrival, the patient's neurological exam was non-focal and there were no clinical concerns or signs of subarachnoid hemorrhage. CT angiography of the head and neck confirmed the basilar tip aneurysm, and also showed the presence of bilateral hypoplastic vertebral arteries and a right-sided PPPIA (Fig. 1). These findings were explained to the patient, and given the history, size, and location of the aneurysm, we recommended treatment. Endovascular intervention was agreed upon, and a stent-assisted coiling procedure was planned. Cerebral angiography revealed a type 1 PPPIA emanating from the right mid-cervical ICA (Fig. 2) that exclusively irrigates the posterior circulation, and demonstrated an irregular wide-necked 10 mm basilar tip aneurysm (Fig. 3).

Given the wide neck of the aneurysm and its incorporation of the left posterior cerebral artery (PCA), we planned to perform stent-assisted coiling. The initial plan was an L-stent, but we were unable to navigate an XT-27 (Boston Scientific Corporation, Fermont, CA, USA) microcatheter into the left PCA given the anatomy of the aneurysm neck. So a Neuroform EZ stent (Boston Scientific Corporation, Fermont, CA, USA) was first placed from the right P1 segment into the distal basilar artery to decrease the neck size. This was not sufficient to provide a scaffold to secure the aneurysm. We were still unable to track an XT-27 (Boston Scientific Corporation, Fermont, CA, USA) microcatheter over the microwire navigated in the distal left PCA. However, we were able to track an SL-10 microcatheter, which allowed us to place a Neuroform Atlas stent (Boston Scientific Corporation, Fermont, CA, USA) from the left P2 into the basilar artery, overlapping the previously-placed Neuroform stent in a crossed Y-stent configuration (Fig. 4). With that, we were able to complete the coil embolization of the aneurysm (Fig. 5).

The patient remained neurologically intact following the procedure



Fig. 2. Right common carotid artery angiography revealing the PPPIA emanating from the mid-cervical ICA taking route of the right vertebral artery.

and was discharged home the next day.

3. Discussion

A persistent primitive proatlantal intersegmental artery is a rare congenital vascular anomaly, its presence signifying an arrest in the embryologic maturation of the vertebrobasilar arterial system. Only approximately forty cases of PPPIA have been reported [6,14–16]. However, within the setting of PPPIA, the presence of additional vascular abnormalities has been reported as frequently as 59% of cases, including multiple reports of aneurysms [13]. We describe the first case of a patient with PPPIA and an unruptured basilar tip aneurysm. Our case is unique in that the PPPIA was the sole vessel supplying the posterior circulation, and therefore the only endovascular corridor through which we treated the aneurysm.

We conducted a search in MEDLINE of the English literature from 1960-current using the following search terms: persistent primitive proatlantal intersegmental artery or PPPIA, and cerebral aneurysm OR intracranial aneurysm. Three anatomical variants of PPPIA have been described, and our case would be classified as Type 1. In Type 1, the origin of the PPPIA is the internal carotid artery, and only two cases of PPPIA Type 1 with a concurrent intracranial aneurysm have been reported [17,18]. A Type 2 PPPIA arises from the external carotid artery, and its occurrence with intracerebral aneurysm has been described in three reports [9,19,20]. Type 3 is the rarest form of PPPIA in which the anomalous vessel originates from the common carotid, and our search did not yield any reports of its co-occurrence with a cerebral aneurysm.

Of the four anastomotic vessels in the developing embryo, the proatlantal intersegmental artery persists the longest until vertebral artery formation. Beginning with the proatlantal intersegmental artery cranially and proceeding to the C6 intersegmental artery caudally, the vertebral arteries form from the transverse anastomoses between the cervical intersegmental arteries. When the proatlantal artery persists, it may be difficult to differentiate from the hypoglossal artery, as both vessels follow a similar course from the extracranial carotid system to the intracranial posterior circulation. However, it is most reliably identified by its entrance into the skull via the foramen magnum, taking a route similar to that of the vertebral artery, versus the hypoglossal

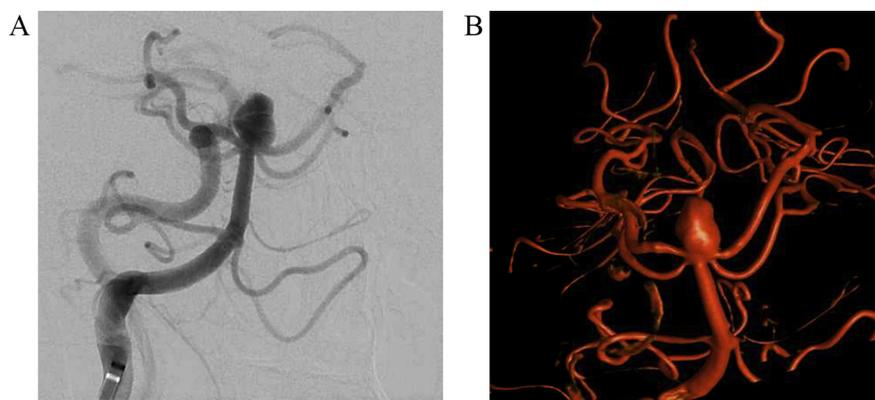


Fig. 3. Right PPPIA angiography demonstrating (A) the wide-necked basilar tip aneurysm, and (B) three-dimensional reconstruction delineating the incorporation of the left PCA origin in the aneurysm.

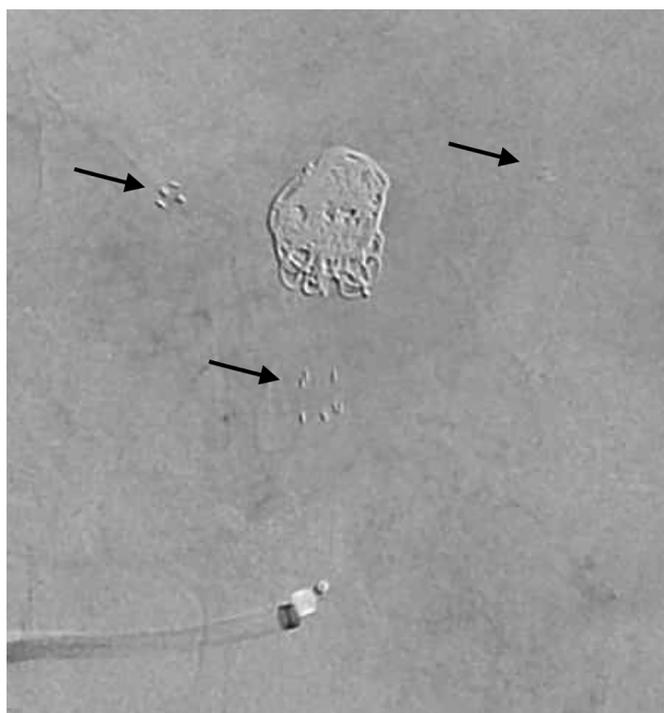


Fig. 4. Skull X-ray demonstrating the Y-stent (arrows) and the coil mass.

artery's entrance into the skull through the hypoglossal canal [1–3].

While some attribute a persistent fetal anastomotic vessel to delayed posterior communicating artery development requiring additional anastomosis to maintain distal basilar flow, others believe the vessel persists in response to a lack of flow in nearby normal vessels. In approximately half of the reported cases of PPPIA, at least one vertebral artery is hypoplastic, if not both, as was seen in our patient [11]. We speculate that the vertebral arteries remained hypoplastic due to the extreme dominance of the PPPIA since the time of its embryologic origination.

The presence of the PPPIA variant limited our endovascular intervention options, especially given the isolated posterior circulation without access through the hypoplastic vertebral arteries and posterior communicating arteries. The angioarchitecture of the aneurysm prevented navigating a standard microcatheter for stent deployment. Ongoing advancement of endovascular tools and the recent release of the Neuroform Atlas stent (Boston Scientific Corporation, Fermont, CA, USA), which is deployable through a smaller malleable microcatheter, allowed for the successful treatment of this aneurysm.



Fig. 5. Right PPPIA angiography at the conclusion of the procedure revealing complete coil embolization of the aneurysm.

4. Conclusion

Persistent primitive carotid-vertebrobasilar anastomosis is often detected incidentally, but can have significant clinical implications. PPPIA is a rare vascular anomaly that is relatively frequently associated with additional intracranial vascular pathology. Awareness and precise understanding of the anatomy of the embryologic remnants is key in planning neuro-interventional access and avoiding complications.

Declaration of Competing Interest

None.

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