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REVIEW PAPER

Proatlantal intersegmental artery: a review of normal and pathological features

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Abstract
Objects Primitive carotid–vertebral and carotid–basilar anastomoses are formed early during human embryogenesis at approximately 24 days. From cephalic to caudal direction, these anastomoses are cranial extensions of the primitive internal carotid, trigeminal, otic, hypoglossal and proatlantal intersegmental arteries.
Materials and methods Normal and/or abnormal morpho-functional aspects of prenatal and postnatal forms of the proatlantal intersegmental artery, from the 24th day of gestation to postnatal eight decades, are described according to personal and literature data. Many (ab) normal carotid–vertebral anastomoses are also marked in differential diagnosis of the proatlantal intersegmental artery.
Conclusions The proatlantal intersegmental artery maintains the posterior circulation until the vertebral arteries are fully developed between the seventh and eighth gestational weeks. When this artery fails to obliterate, it becomes persistent one. The proatlantal intersegmental artery, most commonly, is an incidental finding or it may be of clinical significance in some patients.

Keywords Human artery · Embryo · Fetus · Adult · Carotid-vertebral anastomosis

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Introduction

General embryology of carotid–vertebrobasilar anastomoses

Transitory or primitive carotid–vertebrobasilar anastomoses, as presegmental arteries in the embryonic period, distribute blood from the primitive internal carotid artery (ICA) to the developmental vertebrobasilar system. Until 24–28 days, the primitive ICAs are supplied by the ventral aorta and the third aortic arches. The hindbrain circulation begins to develop from two longitudinal neural plexus that eventually connect in the midline to become the basilar artery (BA). This plexus is initially supplied by the cranial extension of the primitive ICA and below by the cervical intersegmental arteries and four transient anastomotic channels (trigeminal, otic, hypoglossal, proatlantal intersegmental) between the hindbrain vascular plexus and anterior carotid circulation (Fig. 1). In the 5- to 6-mm (28–30 days) embryo, these anastomotic channels provide the proximal supply to the longitudinal neural arteries via the primitive proatlantal intersegmental arteries and cervical carotid–vertebral anastomoses. Involution of these anastomotic channels starts when the embryo is 7 to 12 mm, while the same process in the

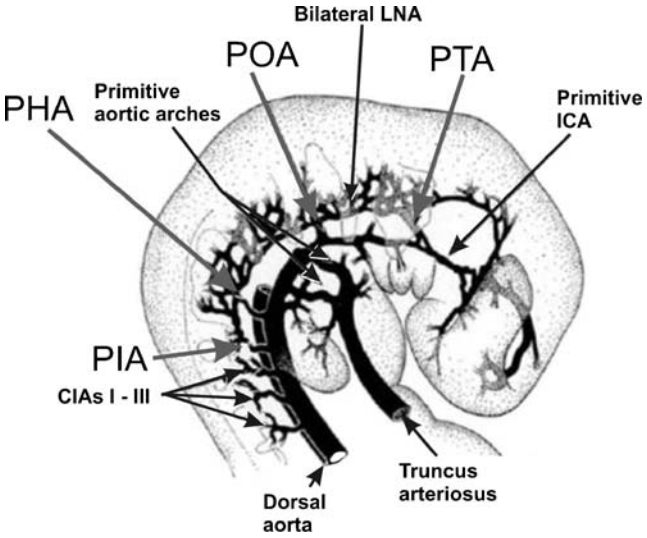

Fig. 1 Modified drawing [14] of cranial arteries at 5 weeks of gestation shows pre-segmental arteries connecting primitive ICA with bilateral LNA plexus. *ICA* internal carotid artery, *LNA* longitudinal neural artery, *CIAs I–III* cervical intersegmental arteries I–III, *PIA* primitive proatlantal intersegmental artery, *PHA* primitive hypoglossal artery, *POA* primitive otic artery, *PTA* primitive trigeminal artery

proatlantal intersegmental artery is the last one [16, 40, 48, 52]. Tsukamoto et al. [75] quoted, according to Congdon [20], that the proatlantal intersegmental artery (PIA) completely disappears on 12 to 14 mm. According to the description of Yilmaz et al. [86], it normally regresses, forming the intracranial vertebral artery (VA) by the end of the sixth gestational week. If the PIA persists on 12 to 14 mm, then it starts from the middle part of the ductus caroticus or near the III primitive aortic arch. Since the ductus caroticus disappears in this stadium, one can suppose that PIA should have the same destiny. Lie [40] quoted, according to Padget [53], that the large PIA stimulates the persistence of the ductus caroticus. After regression of the embryonic trigeminal artery, the PIA and the posterior communicating artery (PCoA) supply the basilar system and make the following carotid–vertebrobasilar communication possible.

Embryonic differentiation

The first cervical intersegmental artery and the primitive hypoglossal artery [54] are important for the differentiation from the PIA regarding the serial numbering of the arteries related to the cervical and occipital embryonic somites (Fig. 2).

Historical data in the literature

In the literature, there are some disagreements about the historical facts. According to Vitums [83], this primitive vessel was already observed and correctly interpreted by Fropier [21] in the bovine embryo and by Hochstetter [30] in the rabbit embryo, as well as by Schmeidel [66] in the human embryo. However, Hutchinson and Miller [34] remarked that in 1885, Gottschau [24] was the first who described this carotid–vertebral anastomosis in human.

According to Padget [54] and her personal communication with Professor Arey, a completely new name, "proatlantal intersegmental artery" became suitable because this artery is located between the occipital and cervical somites; it is accompanied by the first cervical nerve and provides communication between developing carotid and vertebral circulations.

Persistent proatlantal intersegmental artery

The first descriptions

Bloch and Danziger [12] pointed out that the first description of the persistent proatlantal intersegmental artery (PPIA) was

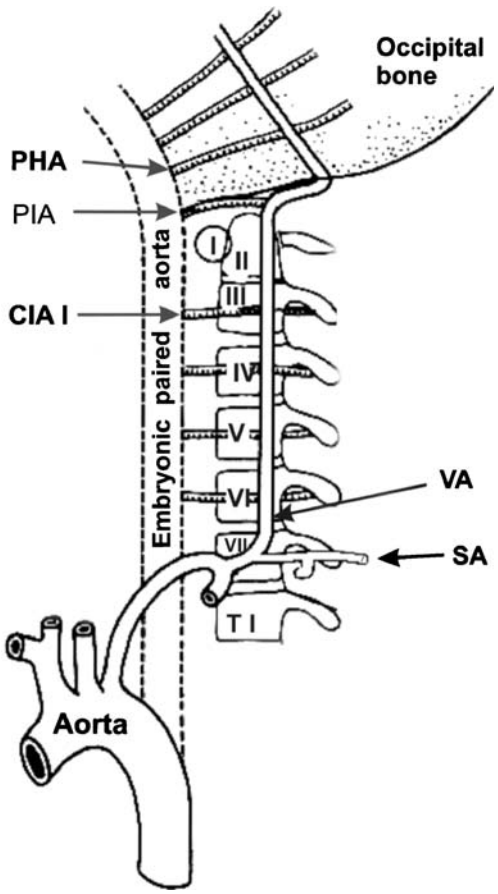

Fig. 2 Modified drawing [54] shows a position of the PIA (primitive proatlantal intersegmental artery) in relation to the PHA (primitive hypoglossal artery) and CIA I (first cervical intersegmental artery). *VA* vertebral artery, *SA* subclavian artery, *I–VII* cervical vertebrae, *T I* first thoracic vertebra

given by Congdon [20], while Pasco et al. [58] attribute its description to Gottschau [24]. The first angiographical description of a PPIA was not given until 1966 [19].

Conforti et al. [19] named this carotid–vertebral anastomosis as the primitive cervical segmental artery; Lasjaunias et al. [39] described it as the proatlantal artery I; Suzuki et al. [71] named it as the suboccipital intersegmental artery, while Pasco et al. [58] named it as the pseudo-vertebral artery.

Rao and Sethi [64] cited the nomenclature used for PPIA from 1885 to 1975 in their Table 1 as follows: (1) abnormal vessel; (2) anomalous carotid–vertebral anastomosis or primitive cervical segmental artery; (3) anomalous origin of vertebral artery from the external carotid artery; (4) external carotid origin of dominant vertebral artery; (5) congenital external carotid–vertebral anastomosis; (6) persistent primitive hypoglossal artery; and (7) proatlantal artery.

Some authors used Latin terms: The arteria intersegmentalis proatlantica [10], arteria proatlantoidea [44], arteria intersegmentalis proatlantis [67], and arteria intersegmentalis proatlantalis [81].

Patterns of beginning, course, and termination

When unilaterally present, PPIA originates from the ICA [14] either at the level of the C2 vertebra [4, 8, 17, 34, 71] or at the level of C2–C3 vertebrae [7, 57] or C3 vertebra [40, 46] or C4 vertebra [12, 50]. However, it was noted only in one case that PPIA begins 2 cm above the origin of the left ICA [25]. Bilaterally, PPIA originated from the ICA [59, 61] at the level of C2 vertebra [27, 84]. Usually, this artery has a horizontal course along the posterior arch of atlas [7, 14, 27] or it shows a strong posterior curve before joining the VA at atlantic part (V3) [58]. Pasco et al. [58] described the common trunk originating from the carotid bulb which gives off two branches: anterior–ICA and posterior so-called pseudo vertebral artery. The persistent proatlantal intersegmental artery is, in its course, more often positioned laterally to the ICA; it makes a sharp curve dorsally and above the atlas entering into the cranial cavity through the foramen magnum [4, 12].

This artery can originate from the external carotid artery (ECA) [3, 6, 11, 32, 41, 44, 60], near the ECA origin [64] or at the level of the atlas, when it has a common origin with the occipital artery [72]. It can also originate from the ECA, either at the level of C2 vertebra [4, 62, 84] or at the level of the C3–C4 intervertebral disc [3] or C4 vertebra [33]. In the case described by Akay et al. [3], the left VA had posterolateral beginning from the ipsilateral ECA at the level of C3–C4 intervertebral disc. The artery then runs posteriorly as a short vascular loop and then ascends to enter the transverse foramen of the atlas. Thereafter, it passes horizontally and posteriorly over the posterior arch of the atlas and entered the intracranial cavity via the foramen magnum.

This artery can originate from the CCA, either unilaterally [55] or bilaterally [41]. An “abnormal artery” that arose from the left common carotid bifurcation and than passed obliquely upwards, between atlas and occiput, with the same course as a normal left VA, was named a PPIA by Palmer and Philips [55].

Types ordering

Type 1

Unilaterally, this type of PPIA [35, 39, 48, 52] begins either from the ECA [4, 41] or the ICA [7, 8, 41, 63] or the CCA [41] (Fig. 3), but it does not pass through the transverse foramina of the cervical vertebrae. It joins with the intracranial part of the VA after the passage through the foramen magnum. Bilateral presence of PPIA-type I was described by some authors [27, 59, 84].

Subtype There is an exception in the description of this type of PPIA. Namely, Schoof et al. [67] discovered the PPIA which originates from the V3 part of the VA and runs towards the ICA distally from the carotid occlusion.

Type 2

The type II of PPIA (Fig. 4) arising from the ECA [3, 6, 32, 39, 44, 48, 62–64] keeps more lateral position than the first type of PPIA and passes through the transverse foramen of C1 vertebra and then joins with the V3 of the VA.

Subtypes The left ECA angiography, described by Ahn et al. [2], showed direct connection between the occipital and left vertebral arteries via the PPIA type II at the level of C1 vertebra. Right and left external carotid angiograms, described by Lui et al. [42], showed two anomalous vessels arising from the proximal part of corresponding occipital artery. One of them, running posteriorly and dorsally, joined the V3 part of corresponding VA.

General remarks

It was postulated that type 1 proatlantal artery is the true PPIA, while type 2 is a persistent primitive first cervical intersegmental artery [52].

Rudiments of the PPIA

Suzuki et al. [71] supported the hypothesis that the occipital artery with the beginning from the ICA represents a rudiment of the PPIA. Furthermore, the rudiments, incorporated into VA trunk [77, 79], caused its partial duplication (Fig. 5).

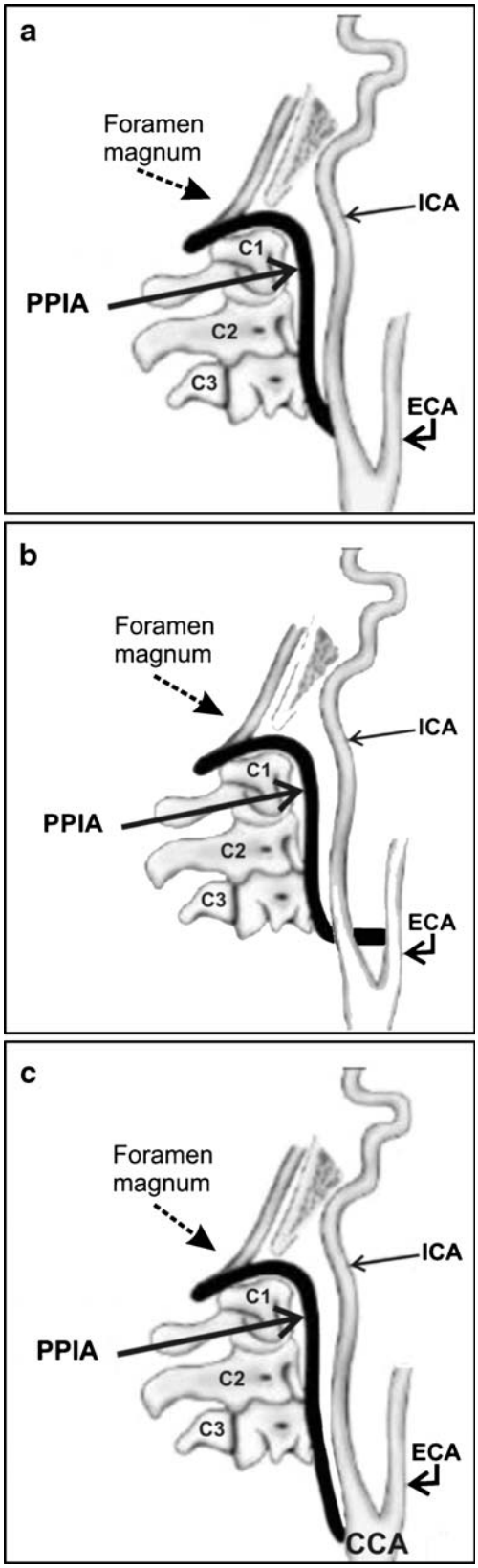

Fig. 3 Modified drawing of the first type of PPIA with its origin either from the internal carotid artery—*ICA* (**a**) or external carotid artery—*ECA* (**b**) [27] or from the common carotid artery—*CCA* (**c**) [55]. *C1–C3* first three cervical vertebrae

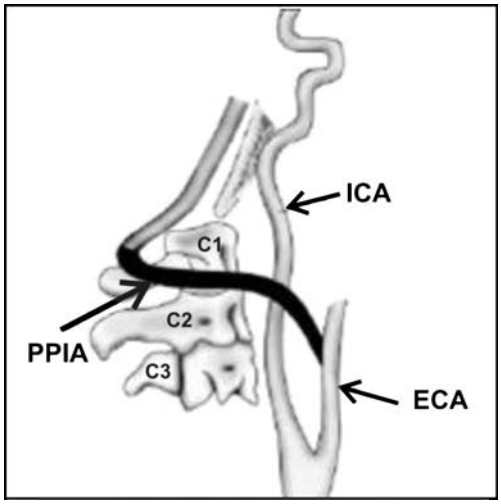

Fig. 4 Modified drawing of the second type of PPIA with its origin from the ECA (external carotid artery) and its passage through foramen transversarium of the C1 vertebra [27]. *ICA* internal carotid artery, *C1–C3* first three cervical vertebrae

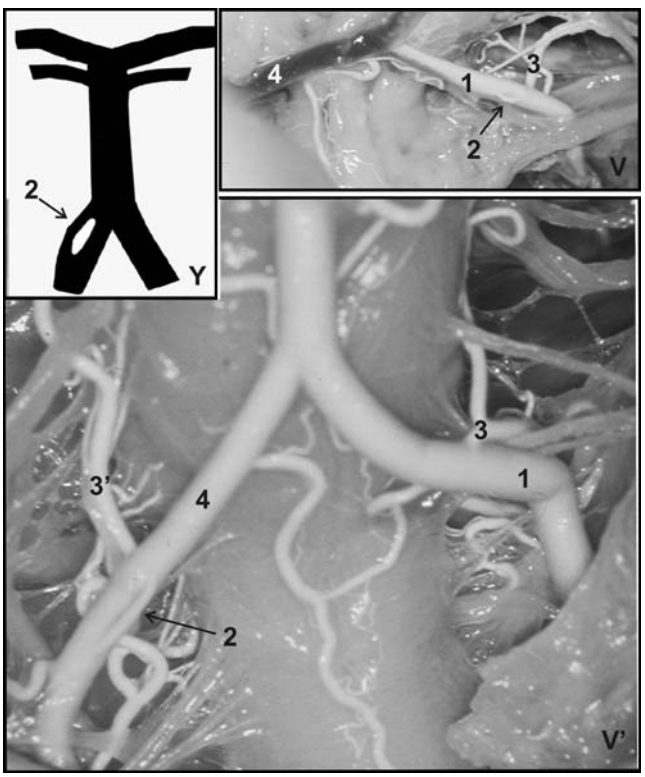

Fig. 5 Composite of the original images (*V* and *V'*) [79] and schema [85] shows status of the VA (vertebral artery) after incorporation of PIA rudiments in its V4 (intracranial) segment. Left VA (*1*); partial persistence of PPIA (*2*); posterior inferior cerebellar artery on the left (*3*) and right (*3'*) sides;
Right VA (*4*)

Caliber

It was described that the diameter of the PPIA can be larger than the diameter of the distal portion of ICA [34] or can have the same size [58] or the diameter with the value of 4 mm [33]. It was described that a sizable PPIA was seen branching from the ECA [64].

When PPIA persists bilaterally, one of vessels can be larger than the second PPIA [27, 84].

PPIA branches

Usually, there are no branches arising from the PPIA. However, the occipital artery can begin from the PPIA [4, 41, 60, 62]. A very rare case was described by Suzuki et al. [71] when the PPIA was divided in two branches above C2 vertebra; one of those branches entered the posterior cranial fosse through the hypoglossal canal and joined with the left VA, and the other one was directed occipitally.

Associated vascular variations

The condition of aortic arch

Basekim et al. [8] discovered a left brachiocephalic trunk of aortic origin which gave the left common carotid and subclavian arteries.

The condition of the VA

Embryologically, it is formed by the fusion of the multiple longitudinal anastomoses between the adjacent cervical intersegmental arteries (from the PIA to the C6 intersegmental artery) [54]. The unsuccessful fusion at any level can result in duplications or fenestrations of the VA [15, 76, 77, 79]. A case of vertebral artery fenestration associated with occipital–vertebral anastomosis was also described [49].

In their summary, Kolbinger et al. [35] concluded that if PIA persists in its entirety, the ipsilateral, contralateral, or both VAs were hypoplastic in 46% of reported cases. An association of the bilateral hypoplastic VA and the left PPIA was confirmed by Bloch and Danziger [12]. On the other side, Bahşi et al. [7] and Basekim et al. [8] discovered a hypoplastic VA on one side and an aplastic VA ipsilaterally with PPIA. In other cases, Gumus et al. [27], Luh et al. [41], and Lui et al. [42] proved bilateral aplasia of the VA. There was also VA aplasia only at prevertebral (V1) and cervical (V2) parts on the side of PPIA [8, 14, 25, 32] or bilaterally [61, 75, 84].

Anderson and Sondheimer [4] proved the presence of normal bilateral VAs, while Akay et al. [3] and Suzuki et al. [71] revealed normal VA on the opposite side of PPIA. The vertebral artery can be also hypoplastic on the opposite side of PPIA and without intracranial connection with the BA [25]. In the presence of the bilateral PPIA and unilateral persistent trigeminal artery, bilaterally tortuous VA was also present at the same time [59].

The condition of the carotid system

The condition of the CCA This artery may be without bifurcation in the ECA and the ICA and the arterial branches that usually emanate from the ECA arose from the CCA directly [8]. Recently, Chan et al. [17] have described the lack of communication between the right CCA and cervical ICA in the presence of PPIA.

The condition of the ECA In the presence of PPIA, the ECA can be absent bilaterally [8]. Horowitz et al. [32] described aortic origin of the ECA with the simultaneous presence of type II proatlantal–vertebral anastomosis.

The condition of the ICA Pascual-Castroviejo and Lopez-Gutierrez [59] described simultaneous kinking of the ICA at two different levels in the presence of bilateral PPIA.

The condition of the PCoA Sometimes, this artery can be unilaterally absent [72].

Association of PPIA with other carotid–vertebrobasilar anastomoses and/or accessory vessels

Simultaneous presence of the PPIA and primitive hypoglossal artery was described by Suzuki et al. [71]. Association of bilateral PPIAs and primitive trigeminal artery in a 6-year-old girl was described by Pascual-Castroviejo and Lopez-Gutierrez [59].

The condition of cerebral veins and dural venous sinuses

In a 6-year-old patient of Purkayastha et al. [62], the bilateral PPIA was associated with an absence of the straight right transverse and both sigmoid sinuses. The deep veins and both internal jugular veins were not opacified, while there was an abnormal falcine sinus connecting the venous sac of the fistula of Galen's vein with the superior sagittal sinus.

Differentiation from other (un)constant carotid–vertebral anastomoses

Persistent primitive hypoglossal artery

A case of a combined anomaly of persistent primitive hypoglossal artery (PPHA) and PPIA was proved on

arteriogram obtained by the insertion of a needle into the common trunk of both arteries [71].

Lie [40] mentioned four essential criteria in establishing the diagnosis of PPHA: (1) the artery arises from ICA at the level between C1 and C3 vertebra; (2) the artery enters the skull through the hypoglossal canal; (3) basilar artery is filled beyond the point where PPHA joins it; and (4) the posterior communicating arteries are absent or not visible on angiograms.

Usually, the PPIA originates at a lower level than PPHA, and it that case, it has a more dorsal sweep compared with more vertical orientation of PPHA [12, 14]. This artery enters the skull via the foramen magnum, the appearance of the atlanto-occipital portion of the vessel differing in no way from that of the normal VA, since the origin is identical in each case [34].

Persistent first cervical intersegmental artery

At the embryologic level, the type I of PPIA is a persistence of the first CIA arising from the dorsal aorta above the ductus caroticus, future second segment of the cervical ICA [58].

Generally, the persistence of one or two cervical intersegmental arteries in adult life is manifested by an abnormal origin or a segmental duplication of the VA [40, 76, 77, 79]. In fact, the absence of any demonstrable anastomosis with the CIAs, coupled with the level of origin from the ICA, effectively distinguishes PPIA from other examples of anomalous development of the VA (persistent CIAs, aortic origin of the VA, etc.) [33].

Carella et al. [15] discovered double extracranial fenestrations of the VA at the C1 and C2 vertebrae levels in a 48-year-old woman, and according to our opinion, they were probably caused by partial persistence of the PIA and the first CIA. Glunčić et al. [23] described an anomalous branch arising from the aortic arch and the serpiginous collection of the blood vessels on the left side of the neck in a 15-year-old girl. These authors considered this aberrant branch as one of the first three cervical intersegmental arteries.

Cervical carotid–vertebral anastomotic canals

Parkinson et al. [56] described an irregular restriction of the supraclinoid part of the left ICA and the short anastomotic canal between the right VA and ICA at the level of C2–C3 vertebrae in a 49-year-old patient with aneurysm of the anterior communicating artery. This anastomotic vessel supplied ICA because there was the absence of the functional connection between the CCA and ICA. These authors noted that this canal should be distinguished from any persistent carotid–vertebral or carotid–basilar anastomosis.

Chan et al. [17] described a case of anastomosis between the VA and ICA at the level of C2 vertebra in a 75-year old patient, as well as an absence of communication between the CCA and cervical ICA on the right side; however, this additional anastomotic channel was more caudal to the PPIA.

In another case, it was described that ICA can be split into ventral and dorsal branches; the ventral branch runs up as the true ICA, whereas the dorsal branch or "pseudo VA" replaces the original left VA. This "pseudo VA" runs straight upwards, almost parallel to the ICA, reaching the C3 transverse foramen where it enters its bony canal and follows the regular course of the true VA [13].

Hacket and Wilson [28] described a large artery which originates from an enlarged right ECA and courses laterally and posteriorly to become the VA. Furthermore, the BA is filled by this artery. The best explanation of this vessel assumes that it represents a congenital abnormality. Several factors can support this opinion: (1) the size of the vessel; (2) the absence of any branches from the vessel; (3) the direct course of the artery opposite to a tortuous channel through branches of the occipital artery; and (4) the marked enlargement of the ECA.

A very interesting operative approach suitable in selected cases of VA insufficiency and subclavian steal syndrome was performed by Clark and Perry [18]. They removed VA in its V2 and V3 parts and positioned it under the sternocleidomastoid muscle, connecting it with the proximal end of the ECA by end-to-end anastomosis.

Variable origin of the occipital artery

Normally, the occipital artery (OA) arises from the posterior aspect of the ECA. However, it can begin from ICA when it is embryologically identical to the beginning of the PPIA [71].

Lasjaunias et al. [39] described that the OA and its different varieties may correspond with the remnant of the first two intersegmental arteries for these reasons: (1) the identical course of the various OAs, their vertebral anastomosis, and two types of PPIA; (2) the varieties of the OA origin from the VA can demonstrate the relationships between the three systems in the human as in the phylogeny; and (3) the apparent segmental distribution of the three muscular trunks and the constancy of the two of them.

Aggarwal et al. [1] noted a variant of origin of the occipital superior thyroid and ascending pharyngeal arteries, arising from a common trunk on the distal end of an occluded ICA, in a 63-year-old male.

Direct or indirect anatomic and/or pathologic carotid–vertebral anastomoses

Occipital–vertebral anastomosis Tsai et al. [74] emphasized the agreement of some authors that the occipital–

vertebrobasilar anastomosis through one or more vascular channels usually represents collateral pathways which have expanded as a result of cerebrovascular disorders. However, these authors noted that the absence of bilateral VAs in their patient was most probably caused by the development of occipital–basilar anastomosis in utero. They also described that if the vascular arrest or occlusion develops at the gestational age of 32–36 days, the BA will exist without the formation of the VA. In order to correspond to the increased metabolic requirements of developing brain and increased pressure gradient, it will change direction of flow via the OA to the BA and lead to a compensatory hypertrophy of the OA.

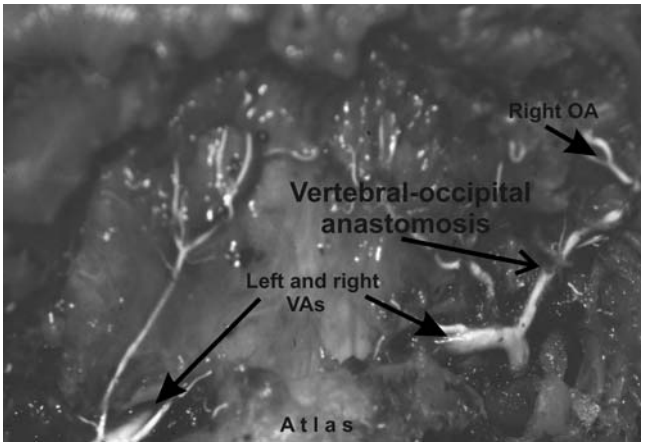

Fig. 6 Original case of the vertebral–occipital anastomosis in the human fetus [76]. There was an anastomosis between the right vertebral artery (*VA*) at atlantic part (*V3*) and occipital artery (*OA*)

The left ECA angiography showed direct connection between the OA and left VA via the PPIA at the level of C1 vertebra [2]. Many years ago, Schechter [65] noted four cases of normal direct vertebral–occipital anastomosis among 1,000 angiograms. However, the author emphasized that there were data in the literature about the persistence of these bilateral direct anastomoses in 45 cases and unilateral anastomosis in five cases (three on the left and two on the right side) among 53 cadavers. Kuwabara et al. [36] performed the catheter cerebral angiography in a consecutive series of 139 patients with various diseases and visualized occipital–vertebral anastomosis in six cases on the left side. The existence of the small but also the permanent anastomotic vessels between the VA and the OA were proven on the cadavers by some researches, as was remarked by Hauke and Zeumer [29]. Although they discovered the anastomosis of this kind only in one case, the previous authors do not share the opinion of Traupe et al. [73] who claimed that collateral connections are opened only in the stenotic processes on the carotid artery as well as on the VA or during the selective angiography. The similar fetal anastomosis between the suboccipital branch of the right VA and ipsilateral OA [76] can be confirmed by the opinion of Traupe et al. [73], since the anastomosis was visible only when the Micropaque was injected under the significant pressure (Fig. 6). Well-developed anastomotic branch, discovered between the occipital and vertebral arteries, can be considered as an anatomical variant of the vascular system [43]. The similar branch in mature persons can also serve as an intermediate link for transmitting irritation at the neck osteochondrosis along the periarterial plexus from the VA to the occipital one which results in reflectory pains in the posterior area of the neck.

Furthermore, there are also some indirect vertebral–occipital anastomoses. The first example of this anastomosis is represented by a case in which a descending branch of the OA in the neck splits into two branches: a superficial one and a deep one; the second one is anastomosed with muscular branches of the VA [69]. Holodny and Newark's study [31] presented a patient whose entire right-sided anterior circulation and most of the posterior circulations were supplied by the ipsilateral ECA via the occipital–vertebral anastomosis through the muscular branches of the second and third intervertebral space.

The second example of indirect vertebral–occipital anastomosis is that one between the branches of the normal OA and some branches of the occipital branch that derives from the VA. Schechter [65] described these anastomoses in vertebral angiograms in 50% of the cases. The third example is an anastomosis between the meningeal branches of the VA and the OA in the posterior cranial fossa [37]. Hauke and Zeumer [29] claim that there is an anastomosis between the ECA and VA described on cadavers in most of cases. These authors discovered a case of a large opened anastomosis among five cases of anastomosis between the ECA and VA; in this case there was not the pathoanatomic background in any of these two systems. But in the other four patients, they proved anastomoses between the ECA and VA which were caused by the stenosis in one or another arterial system.

It is well known that the occlusion of an artery is often accompanied by the development of collateral circulation through the dilatation of existing anastomosis. With occlusion of the CCA or the beginning part of ECA, the flow in the ipsilateral ECA and sometimes in the ipsilateral ICA can occur mainly from the ipsilateral VA to the ECA through its occipital branch. Selective injection of the VA ipsilaterally to the occluded CCA with delayed images reveled a retrograde contrast filling of the occipital branch of the ECA through carotid–vertebral collateral [51].

From the clinical point of view, Moret et al. [47] successfully carried out the balloon occlusion of congenital

arteriovenous malformation of the VA, thanks to the existence of the vertebral–occipital anastomosis.

Ascending pharyngeal–vertebral anastomosis An anastomosis between the meningeal branches of the vertebral and ascending pharyngeal arteries is also indicated in the study of Lasjaunias and Moret [38] and in the Gray's anatomy book [69]. Especially, an anomalous vessel that originated from the left ECA and that ascended vertically along the posterolateral wall of the pharynx and then descended tortuously terminating in the left VA at a level of C2–C3 intervertebral disc was described by Shimamura et al. [68]. It was the first documented case of ascending pharyngeal–vertebral anastomosis associated with the bilateral absence of VAs at their origins.

Posterior meningeal artery Usually, it arises from the VA. In some cases, it can originate from the ICA at the level of C2 vertebra, and after the suboccipital course, it passes through the foramen magnum and then divides in the tentorium and falx cerebelli. Except on the beginning part and during the course, the posterior meningeal artery did not have any morphological characteristics of PPIA. In the same article, it was remarked that a similar case was described in the Pernkopf's anatomy atlas [37].

Stylomastoid artery Grobovschek [26] quoted the comments from the doctoral thesis of his colleague, about the stylomastoid artery, which is interesting from the embryological and the anatomical point of view. He emphasized that its communication with carotid and vertebrobasilar systems also represents their arterial anastomosis.

Posterior communicating–vertebral anastomosis The description of an indirect anastomosis of the PCoA and VA, in the embryological and the anatomical point of view, is the unique case in the literature. This refers to the case of a 12-year-old patient with aneurysm of the BA, simultaneously with dilated BA and VA [45]. Meyer et al. [45] angiographically confirmed the reflux of the blood from the patent left PCoA to the ipsilateral VA and explained that it was caused by the presence of some collateral between these two arteries. However, in a 40-year-old man, the midsegmental BA aplasia vas associated with a tortuous right PCoA, which was the major feeding vessel to the posterior cerebral and superior cerebellar arteries as well as to the distal BA [9]. In a 73-year-old patient of Shimamura et al. [68], the vertebrobasilar system was fed via the right PCoA and the anomalous anastomosis from the left ECA. On the other side, Chan et al. [17] proved the supply of the middle cerebral artery by the vertebrobasilar circulation through PCoA in a case of ipsilateral PPIA.

Morphofunctional relevance

The evolution of the PIA is not as clear as the evolution of the primitive trigeminal, otic, and hypoglossal arteries. In fact, the PIA is thought by Behari et al. [9], Bloch and Danziger [12], and Padget [54] to contribute to the V3 part of the VA.

Sometimes, the carotid–vertebrobasilar anastomoses of small caliber cannot be visible on an angiogram [9]. The persistence of carotid–vertebrobasilar anastomoses is not only associated with an altered blood flow between the anterior and posterior cranial circulations but also leads to a suppression of the normal development of the cerebrovasculature [67]. This explains that an ipsilateral or bilateral hypoplasia [4, 42] of the VA is often observed in patients with PPIA. Furthermore, PIA can persist for the reason of the both VAs aplasia. Therefore, the influx of blood into the posterior cranial fossa is possible, thanks to PPIA, and if a surgical treatment is recommended, the integrity of its vascular source has to be maintained [32, 75]. An additional hypoplasia of the right anterior cerebral artery made an anterior cross-flow impossible and led to the critical situation in which the entire left hemisphere was supplied only by the PPIA [67].

According to certain authors, the finding of the PPIA is an incidental finding; however, it may be of clinical significance in some situations [12, 27, 55, 58]. This artery mainly has a contact with the first cervical nerve which potential compression has no clinical manifestation [58]. This artery may account for unusual symptoms, particularly if it is bilaterally presented [84]. Although the PPIA itself was of no clinical significance in some case, its recognition is important if surgical ligation or embolization of the ECA is planned [42]. Only the hypoglossal artery and PPIA connect the carotid system with the vertebrobasilar system in the neck and could be encountered during cervical dissection. Surgical dissection in this region, especially when performing a far lateral transcondylar approach to the foramen magnum and to the cervicomedullary area, can discover the presence of PPIA [22].

Very interesting was the finding of anomalous origin of the left VA from the thyrocervical trunk in a 36-year-old woman whose father was known to have bilateral PPIAs with absence of the VAs. Previous observation and the finding of vertebral anomalies in the patient's father lead authors to hypothesize that those vascular variations may be hereditary [70].

The literature does report a left-sided predilection of PPIA [4, 14, 35, 42]. The youngest patient with a presence of the PPIA was a 3-day-old male neonate [62]; nevertheless, there are more evidenced PPIAs in adults over 40 years of age. Different diseases—headache [2, 12, 34, 42, 50], vertigo [3, 8, 41], syncopal episode [4], parenchymal

hemorrhage [5], epistaxis [6], top of basilar syndrome [7], intracranial parenchymal arteriovenous malformation [11], sudden collapse [17], right-sided paresis [25], left-sided weakness [27, 28], transient ischemic events [32], dizziness [41], symptoms of vertebrobasilar ischemia [44], motor vehicle accident [55], subarachnoid hemorrhage [46, 58], cutaneous hemangioma [59], seizure [60], chronic limb ischemia [61], history of rapidly enlarging head size and gradual proptosis of both eyes [62], deterioration of vision [64], acute right-sided sensorimotor hemiparesis [67], hydrocephalus [71], and stroke [72] were the reasons for the evidence of the PPIA existence. Babu Manohar et al. [6] pointed out a carotid–vertebral anastomosis as the cause of the uncontrolled epistaxis in a patient, allowing appropriate ligation of the patent artery. The case of Palmer and Philips [55] is unusual because of PPIA origin from the common carotid bifurcation. This is the most common site of atheromatous disease of the extracranial arteries. An atheromatous lesion at this site can produce ischemia in both carotid and vertebral territories. Atherosclerotic plaques at the proximal portion of both ICAs and right PPIA were the source of the emboli and consequent neurological symptoms described in the paper of Gumus et al. [27]. In the case of Bahşi et al. [7], emboli rose from the left ICA and passed to the vertebrobasilar system through PPIA and occluded the thalamoperforating arteries, resulting in "top of the basilar syndrome".

In the case of Grego et al. [25], the left ICA, as a source of PPIA, had a 60% intracranial stenosis, while there was a 40% right carotid bifurcation stenosis with a 50% intracranial tandem stenotic lesion. Therefore, as previous authors quoted, when primitive carotid–basilar anastomoses are present, the carotid bifurcation stenosis may be responsible for the symptoms in both anterior and posterior cerebral territories, especially when an incomplete CAC coexists.

The association of basilar bifurcation aneurysm and PPIA has not been presented in the literature until 2001 [50]. The frequency of PPIA combined with the intracranial aneurysm is relatively high, whereas the occurrence of PPIA is extremely rare.

Conclusion

The coronal view of magnetic resonance angiography by phase contrast method is useful for the diagnosis of PPIA: (1) The proatlantal intersegmental artery originates either from the CCA bifurcation or from the ECA or ICA; (2) the level of its origin ranged from C2 to C4 levels; (3) it joins the VA in the suboccipital region; and (4) it traverses the foramen magnum [41].

There were less number of authors who discovered PPIA in children up to 6 years of age [62, 72], while there are more evidenced PPIAs in adults over 40 years of age. A large angiographic study of 4,400 patients revealed the occurrence of primitive carotid–basilar and carotid–vertebral anastomoses in 0.14% and 0.023%, respectively [86]. Quijano et al. [63] noted that 57% of the described PPIAs are of the type 2 variety (origin from ECA), 38% are type 1 (origin from ICA), and 5% of PPIAs arise from the CCA.

A 59% incidence of cerebrovascular abnormalities has been reported in patients with PPIA, 10% of whom had intracranial aneurysms [35]. Therefore, it was suggested that some congenital and/or hemodynamic factors changed by PPIA may affect the pathogenesis of intracranial aneurysms. The frequency of PPIA combined with the intracranial aneurysms is relatively high, whereas the occurrence of PPIA alone is very rare [50].

As the multislice computed tomography angiography is being used more frequently in routine clinical practice, one would more often come across these persistent vessels [3].

The author is aware of morphofunctional and clinical significance of the carotid–vertebrobasilar anastomoses [77–81], as in the case of the hypoglossal artery [82], as well as in the proatlantal intersegmental artery, which is now represented. The review of the other carotid–basilar anastomoses will be done in future manuscripts.

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