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#### ARTICLE INFO

Article history: Received 6 May 2024 Accepted 15 July 2024

Keywords: Proatlantal artery Trigeminal artery Carotid-vertebrobasilar anastomosis Vascular embryology

#### ABSTRACT

Transient embryonic carotid-vertebrobasilar anastomoses can intermittently persist beyond the embryonic period. These vascular anomalies are often serendipitously identified during evaluation for unrelated disease states and pathologies. The persistent proatlantal intersegmental artery is one such recognized rare fetal anastomotic connection, often arising unilaterally. Bilateral persistence of the proatlantal anastomosis is a rarer occurrence, seldom described in the literature. We report a case of bilateral concurrent type I and type II proatlantal arteries, describe the embryology of persistent carotid-vertebrobasilar anastomoses, and consider pertinent clinical implications.

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### Introduction

The persistent primitive proatlantal intersegmental artery represents a rare variant of the fetal carotid-vertebrobasilar anastomoses; and, similar to other persistent fetal anastomotic channels, it represents involutional failure of embryonic vasculature during early development [1,2]. Two variants of proatlantal arteries have been described, arising from either the internal or external carotid vasculature [3–5]. The type I proatlantal artery, described in previous literature as the proatlantal intersegmental artery, arises from the internal carotid artery and ascends to the atlantooccipital region and constitutes the distal ipsilateral vertebral artery which then extends intracranially through the foramen magnum. The type II proatlantal artery, described previously as the first cervical intersegmental artery, arises laterally from the external carotid artery, and passes the C1 or C2 vertebra obliquely to continue as, or perfuse, the distal portion of the vertebral artery, before similarly entering the foramen magnum. Reported cases of a persistent proatlantal artery are infrequent, with unilateral involvement more common [6]. Bilateral persistent proatlantal anastomoses are exceptionally rare and seldom described in the radiologic anatomical literature, and when so, have identical vascular origins from either the internal or external carotid artery on both sides. In the present case, we describe an instance of bilateral concurrent type I and type II persistent fetal proatlantal anastomoses.

\* Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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https://doi.org/10.1016/j.radcr.2024.07.082



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Fig. 1 – Reconstructed images from a computed tomography angiogram, sagittal (A–C) and coronal (D) projections. The origin of a type I proatlantal artery from the right internal carotid artery at C3 can be seen (*arrow*). The anomalous vessel courses superiorly and then with distinctive suboccipital horizontal course (B–C) before constituting the ipsilateral vertebral artery (D).

#### Case report

A 27-year-old male was brought by air to the emergency department following a motor vehicle collision. Computed tomography (CT) of the head and neck was performed which demonstrated a frontotemporal calvarial fracture, subdural hemorrhage, cerebral contusions, nondisplaced bilateral first rib fractures and C7 spinous process and laminar fractures. Cervical angiogram imaging was performed to identify arterial vascular injury. No traumatic injury to the cervical vasculature was identified. The distal extradural and intracranial segments of the vertebral arteries were constituted by vessels arising from the internal carotid artery on the right and the external carotid artery on the left. More specifically, the anastomotic vessel on the right originated from the internal carotid artery at the level of the C3 vertebra and it ascended superiorly to the C1 transverse process and coursed horizontally within the suboccipital region (see Fig. 1). An anomalous vessel was also identified arising from the external carotid artery on the left at the C3-C4 interspace while extending superomedially and passing the C1 vertebra transversely to perfuse the distal V3 and V4 segments of the vertebral artery (see Fig. 2). The occipital arteries arose from the anomalous anastomotic vessels on both sides. The proximal segments of the vertebral arteries were developmentally hypoplastic with corresponding hypoplasia of the bilateral transverse foramina in these regions. Findings were consistent with bilateral persistent proatlantal fetal anastomoses, with type I on the right and type II on the left. No additional aberrant vascular connections between the carotid and vertebrobasilar system were present. No other vascular anomalies or aneurysms were identified.

#### Discussion

During early development when embryonic length is 4 to 5 mm, the primitive vertebrobasilar system receives transient vascular supply from two parallel neural arterial plexuses constituted by the carotid system from trigeminal, hypoglossal, otic, and proatlantal arteries [5–9]. The intracranial and presegmental anastomotic arteries are named according to the cranial nerves with which they course, or with their relationship with the rudimental otic vesicle. The caudal portion of the neural arterial plexus is perfused by eight extracranial anastomosing segments which become intersegmental with development of the cervical vertebral bodies [2,6,8,9]. The first intersegmental artery is named according to its association with the proatlas, a rudimentary vertebral element that arises between the atlas and occipital bone, and supplies a caudal portion of the neural arterial plexus. Seven additional cervical intersegmental arteries, which accompany the corresponding nerve roots, arise from the dorsal aorta and perfuse the caudal end of both neural arteries. The anastomotic channels between the carotid and vertebrobasilar system are generally patent for a period of 7 to 10 days [9]. With maturation and development of the posterior vascular channels, the otic, hypoglossal, and trigeminal anastomoses involute at the 7 to 12 mm embryonic stage in sequential manner, with the proatlantal intersegmental artery persisting until the vertebral ar-



Fig. 2 – Computed tomography angiogram, sagittal (A–F) projection. The origin of a type II proatlantal artery from the proximal left external carotid artery at C3-C4 is seen (*arrow*) in (A–C). The vessel courses superiorly and traverses the C1 vertebra obliquely before constituting the distal left vertebral artery (D–F).

teries develop by the 12 to 14 mm embryonic stage [7,9,10,11]. Six (C1–C6) cervical intersegmental arteries anastomose longitudinally to form the vertebral artery while the distal vertebral suboccipital segment is formed by the distal proatlantal artery [2,5,6]. The seventh cervical segment (C7) is augmented and forms the subclavian artery.

Persistent anastomotic connections between the carotid and vertebrobasilar system are rare, with order of increasing frequency inversely related to presegmental involutional order. Persistent unilateral intersegmental proatlantal anastomosis is exceptionally rare with an estimated incidence of 0.33% [12]. No published data is available to our knowledge regarding the incidence of bilateral proatlantal anastomoses. The proatlantal artery is characterized by a vascular channel arising from either from the cervical internal or external carotid artery which ascends and constitutes the distal transverse suboccipital segment of the vertebral artery near the foramen magnum [2]. It has been suggested that the type I proatlantal artery represents the true persistent proatlantal intersegmental artery while the type II proatlantal artery may represent a persistent primitive first cervical intersegmental artery [8]. No branches typically arise from the proatlantal vessel; however, in some instances, similar to the presented case, the occipital artery originates from the proatlantal artery [5,10]. The vertebral artery segments proximal to the proatlantal anastomosis are generally absent or developmentally hypoplastic [13,14].

Discrimination between the type I proatlantal artery from the more frequently encountered hypoglossal artery necessitates some scrutiny in view of their similar vascular origins from the cervical internal carotid artery. The proatlantal artery originates at a lower cervical level, typically at the C2 or C3 vertebra, while the hypoglossal vessel originates at the C1 vertebra or C1-C2 interspace and has a more anterior course before entering the posterior fossa through the ipsilateral hypoglossal canal [7,15]. The primitive hypoglossal artery also lacks the suboccipital horizontal course which is distinctive of the proatlantal artery.

The present literature suggests that persistence of the trigeminal artery into adult life is more frequent than the hypoglossal or proatlantal anastomoses, with estimated incidence of 0.03% to 2.2% [4,7,9]. The trigeminal artery represents a vascular channel from the proximal cavernous portion of the internal carotid artery to the distal third segment of the basilar artery [7,9]. The persistent otic artery arises from the petrous internal carotid artery, traverses the internal auditory canal, and joins the caudal segment of the basilar artery [11]. There is controversy regarding the otic artery, with some authors recently disputing its persistence into adulthood; nonetheless, the otic artery is felt to have only a minor contribution to the developing rudimentary vertebrobasilar system [4,7,11]. Reported cases of a persistent proatlantal artery are infrequent, with bilateral involvement a rarer occurrence. Previously described bilateral persistent proatlantal anastomoses have identical vascular origins on both sides from either the internal or external carotid artery [6,10,14–17]. To our knowledge, this is the first reported case with concurrent bilateral type I and type II persistent proatlantal arteries. It has been theorized that persistence of these vascular channels may be related to a compensatory effort to maintain adequate perfusion to the posterior circulation during periods of dynamic cerebral development or with developmental failure of the vertebral artery [12,15]. However, the precise etiology and contributing factors to involutional failure are still unclear [9].

While most persistent carotid-vertebrobasilar anastomoses, including the proatlantal artery, are incidentally found, there are procedural and clinical implications related to changes in hemodynamics and cerebral blood flow. The persistence of fetal vascular anastomoses is particularly salient for neurointerventional and endovascular planning and may necessitate procedural modification if present. Similarly, a persistent proatlantal artery heightens risk for iatrogenic injury during carotid endarterectomy, upper cervical fixations, and external carotid ligations [6,11]. Atheromatous disease of the carotid bifurcation or occlusion of the carotid system may induce symptoms of posterior fossa ischemia due to changes in cerebral blood flow. Several cerebrovascular abnormalities associated with a persistent proatlantal artery have been described including vertebral artery aplasia or hypoplasia, additional carotid-vertebrobasilar anastomoses, arteriovenous malformations, and vein of Galen malformations [5,6,10,12]. While the proatlantal artery is also seen in association with aneurysms involving the circle of Willis vasculature, no aneurysms involving the anastomotic proatlantal vessel itself have been described. Moreover, no consensus exists whether this represents a true association or is simply incidental; although, it is postulated that hemodynamic changes associated with persistent fetal anastomoses may partially influence the pathogenesis of intracranial aneurysms [5,11]. A causal relationship has also been demonstrated between the proatlantal artery and pulsatile tinnitus, primarily related to changes in regional blood flow and increased turbulence [3]. Headache, vertigo, visual disturbances, and history of rapidly enlarging head circumference have also been attributed to a persistent proatlantal artery in the literature [5,10,13–15].

## Conclusion

Persistent anastomotic remnants of the fetal carotidvertebrobasilar circulation, such as the proatlantal artery, are infrequently described. Bilateral persistence of the proatlantal vessel is a rarer occurrence and ,when described, have identical vascular origins from either the internal or external carotid vessels bilaterally. The current case describes bilateral concurrent type I and type II proatlantal arteries which were incidentally found. Findings are salient for diagnostic radiologists and interventionalists in view of their clinical and procedural implications.

## Patient consent

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Consent for publication has been obtained from the patient.