

Duplication of the distal end of the left vertebral artery with fenestration of the right posterior cerebral artery

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Abstract

The vertebrobasilar system (VBS) consists in the intracranial parts of the vertebral arteries (VAs), the basilar artery (BA) and its branches. The presence of a duplication at the level of the intracranial segment of VA (V4) is generally an incidental finding, but may be associated with aneurysms or arteriovenous malformations. We present an extremely rare case of duplication of the distal end of the left vertebral artery, associated with fenestration of the right posterior cerebral artery. The distal end of the left VA was duplicated into two arms (the right with a length of 5.5 mm and a diameter of 2.3 mm that connected with the contralateral VA; and the left with a length of 11.0 mm and a diameter of 1.6 mm, which connected more distally with the BA). The right posterior cerebral artery (PCA) had a fenestration in the posterior segment of the posterior communicating part (P2), with a length of 6.8 mm.

Keywords: vertebral artery, duplication of the distal end, posterior cerebral artery, fenestration.

Introduction

The vertebrobasilar system (VBS) consists in the intracranial parts of the vertebral arteries (VAs), the basilar artery (BA), and its branches. The VBS provides blood supply to the medial portions of the temporal lobe, lateral and medial sides of the occipital lobe, cerebellum, brain stem and spinal cord [1]. Anatomical variations of the cerebral arterial system frequently occur due to the complexity of the developmental process. The anatomical variations of the cerebral circulation include: duplications, fenestrations, persistent fetal arteries, and hypoplasia or aplasia of the arteries [1–7]. The prevalence of VA fenestrations is reported as 0.23–1.95% in angiographies and autopsy studies [8]. Intracranial VA fenestrations, determined by magnetic resonance (MR) angiographic studies, were observed in 0.54% of cases [3]. Bergman *et al.* [9] reported that unilateral duplication was much rarer. In 0.72% of studied cadavers, more than two-thirds were found in the extracranial segment of the VA. Additionally, posterior cerebral artery (PCA) fenestrations are extremely uncommon, with a prevalence of 0.05% [2].

We describe a case of duplication of the distal end of the left VA, with associated fenestration of the right PCA.

Case report

Our group reports a case of a 9-year-old boy with no significant past medical history who presented to the

emergency department with diplopia, headache, neck stiffness, left oculomotor nerve palsy, and nausea, that started 24 hours prior to admission. The patient suffered a traumatic head injury two weeks prior to control. Computed tomography (CT) scan of the head showed a moderate subarachnoid hemorrhage in the posterior fossa, secondary to bleeding of the lower-arm of the right PCA fenestration.

MR angiography was performed at the Neuromed Diagnostic Imaging Center (Timișoara, Romania) using a 1.5-Tesla MR scanner. It revealed diffuse edematous changes of the retrobulbar fat with discrete signal abnormalities that showed a hypointense signal of the left optic nerve. Furthermore, two extremely rare anatomic variations were observed: a duplication of the distal end of the left VA with dominance (larger diameter) of the right VA (Figure 1, A and B), and a fenestration of the right PCA (Figure 1C). Measurements of the length and diameter were performed using multi-planar reconstruction (MPR) technique. Continuity between the arterial lumens at the level of the fenestrations was performed using shaded surface display (SSD) (virtual arterial endoscopy).

MR angiography examination demonstrated a tortuous path of the VAs with predominance of the right VA (right/left VAs diameters in the proximal part of V4 segment was 4.1/3.0 mm). The distal end of the left VA is duplicated into two arms (the right with a length of 5.5 mm and a diameter of 2.3 mm that connected with the contralateral VA; and the left with a length of 11.0 mm

and a diameter of 1.6 mm, which connected more distally with the BA). The distance between the connection points (right vertebral artery and basilar artery) of the right and left arms, respectively, was 8.5 mm. The BA had a

diameter of 3.7 mm and a length of 19.3 mm. The right PCA had a fenestration in the posterior segment of the posterior communicating part (P2), with a length of 6.8 mm.

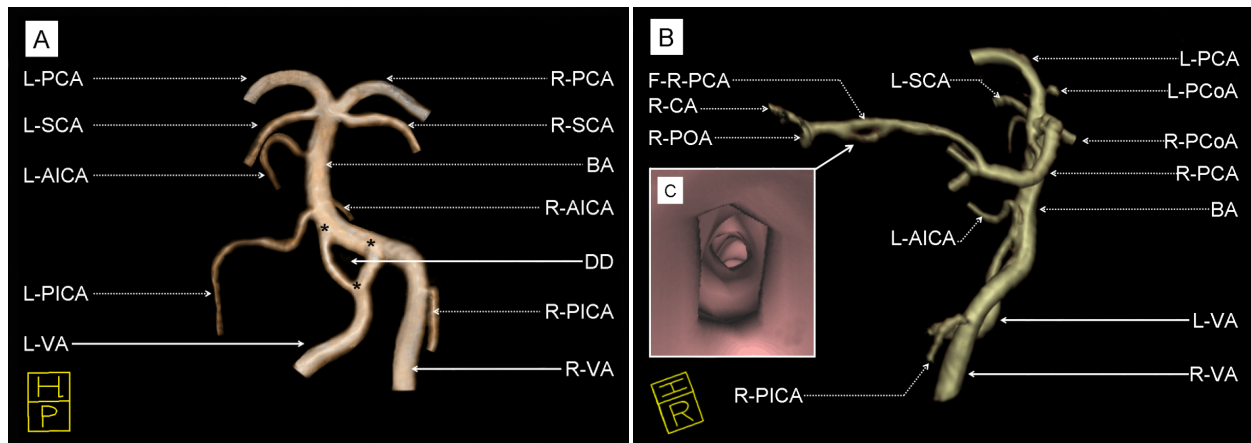


Figure 1 – Three-dimensional MDCT angiography reconstruction of the skull base arteries. Volume rendering technique (VRT) images of vertebrobasilar system with emphasis on a vertebrobasilar junction duplication and a fenestration of the right posterior cerebral artery. (A) Postero-superior 3D image of the vertebrobasilar system. The superior cerebellar arteries (SCA) originate symmetrically from the upper end of the BA. The left AICA originates from the middle third of the BA. The left PICA and right AICA originates symmetrically from the lower third of the BA. The right PICA originates from the distal segment of the right VA. (B) Right-superior 3D image of the basilar artery and the right posterior cerebral artery. The right PCA had a fenestration in the posterior segment of the posterior communicating part. (C) Coronal shaded surface display (SSD) (virtual arterial endoscopy) image indicating the lower arm fenestration of the distal third of the right PCA. L: Left; R: Right; VA: Vertebral artery; DD: Distal duplication; BA: Basilar artery; PICA: Posterior inferior cerebellar artery; AICA: Anterior inferior cerebellar artery; SCA: Superior cerebellar artery; PCoA: Posterior communicating artery; PCA: Posterior cerebral artery; F: Fenestration; CA: Calcarine artery; POA: Parieto-occipital artery.

The intraluminal diameters of the fenestration arms was measured (in coronal plane) at the level of proximal, middle and distal thirds. At these levels, the upper arm had a diameter (height/width) of 1.8/1.5 mm, 1.7/0.9 mm, and 1.7/0.8 mm, respectively. The lower arm had a diameter of 1.3/1.1 mm, 0.9/0.8 mm and 0.8/0.5 mm, respectively. The distal third of the lower arm, the bleeding area, had the smallest intra-luminal diameter of the fenestration arms. The left anterior inferior cerebellar artery (AICA) originated from the middle third of the BA, and the right AICA originated from the distal third of the BA. The left posterior inferior cerebellar artery (PICA) originated from the lower third of the BA, and the right PICA from the distal third of V4 segment of the right VA.

After an extensive evaluation of the child's condition, he was subjected to a specialized endovascular treatment center for coiling of the lower-arm of the right PCA fenestration.

Discussion

The VA is divided into four anatomical parts: (V1) the origin to its entry into the transverse foramen of C6; (V2) the transverse foramen of C6 to C1; (V3) the exit from the transverse foramen of C1 to the foramen magnum; and (V4) the foramen magnum to the BA. The first three anatomical parts are located extracranially, while the fourth is located intracranially [8]. Malformations of the VA may include: (i) duplication and fenestration, (ii) anomalies of width and length, (iii) convolutions, (iv) abnormal variations in course and dural crossing, (v)

collateral branch variations, and (vi) persistence of fetal arteries [9].

The difference between arterial fenestration and duplication must be noted. In fenestration, the vessel has a single origin. The trunk then divides into the vessel into two lumens along its course, which may or may not have a common tunica adventitia. In contrast, a duplication consists of two independent vessels that remain separate throughout their course [1]. Polguy *et al.* [4] also adds that there is fusion of duplicate arteries at the neck. An analysis of the literature [1] revealed that there are alternative names used for 'fenestration' and 'duplication'. 'Fenestration' is otherwise referred to as 'segmental duplication' or 'partial duplication', and 'duplication' is also termed 'extreme fenestration'. Proximal segment BA fenestrations have been described as having a small slit like configuration [3]. This condition was similarly described by Kovač *et al.* [7]. Lasjaunias *et al.* [10] reported on premedullary duplication of a VA as an enlarged premedullary bridge.

According to Weis *et al.* [11] duplication of a VA is a rare congenital anomaly that can be because of the failure of regression of primitive vessels during embryological development occurring between the 32nd and 40th day. Fenestrations of the VBS are due to failure of the complete fusion of the two neural arteries of the embryo (between the 29th and 30th day of fetal development) [2] and intracranial VA fenestrations may form as the result of persistence of the anterior arterial collateral system [10, 12].

Fenestrations and duplications of the cerebral arteries can be associated with: cerebral aneurysms [2], another

(multiple) fenestration [2], variations in the vessels [1], persistent carotid-basilar anastomoses [7] arteriovenous malformations [1], and differences in the relationship of nerves with the cerebral vessels [2]. In our case, there was a fenestration of the right PCA in association with duplication of the distal end of the VA.

According to Bergman *et al.* [9], unilateral duplication of the VA was reported in 0.72% of studied cadavers, and one-third was situated in the extracranial segment of the VA. Bayrak *et al.* [2] reported in 395 patients that fenestrations of the cerebral arteries had an incidence of 7.39%. Fenestration of the anterior communicating artery represented 5.32% of cases, the middle cerebral artery, 1.01% of cases, and the superior cerebellar artery, 1.01% of cases. PCA fenestrations were rare, usually occurring at the proximal portion of the arterial trunk, and had a reported incidence of 0.05%.

Asymmetry of the origin of AICA and PICA is well documented in the literature [1, 7, 10, 13], and has a well-founded embryological explanation with the basis of failure of fusion. In our case, the symmetry of the origin of the AICA and the PICA correlated with the tortuous course of the VAs and the BA, which deviated to the right.

The complex anatomy of the vertebrobasilar junction (VBJ) and the proximal BA account for the prevalence of fenestrations and duplications make this region difficult to approach surgically [14, 15]. Alternatively, VBJ aneurysms can be managed *via* endovascular embolization with Guglielmi detachable coils [7].

☒ Conclusions

Duplication of the distal end of the VA is an extremely rare anatomical variation. It is an incidental finding, but may also be associated with aneurysms or arteriovenous malformations. We report for the first time the presence of fenestration of the right PCA, in association with duplication of the left VA. It is crucial that prior to surgical or radiological cerebrovascular intervention, such anatomical variations of the cerebral arteries to be identified.

Conflict of interests

The authors declare that they have no conflict of interests.

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Received: May 14, 2014

Accepted: April 22, 2015