

Nonaneurysmal Cerebrovascular Conditions Associated with Intracranial Arterial Fenestrations

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Abstract

Background: Cerebral arteries have been identified with multiple anatomical variations, including a concomitant fenestration with aneurysms, but no association is proven. **Aim:** to investigate non aneurysmal disorders associated with intracranial arterial fenestration. **Methods:** A thorough and detailed analysis of the available literature from 1970 to 2020 in PubMed were contemplated to identify and address all the disorders associated with arterial fenestration with exclusion of intracranial aneurysms. **Results:** While segmental vulnerability may induce invisible anatomical histological and hemodynamic changes, cadaveric studies showed that the frequency of fenestration is up to 40% higher than the clinical reports, and the cadaveric reports also showed a higher occurrence of such fenestrations as compared to radiological studies. The vertebrobasilar system, the most common site of fenestration. Fused vertebrae and other vascular defects of up to 7% are previously associated with the vertebral artery fenestration. **Conclusion:** intracranial fenestration is a critical anatomical variant. Thus, A comprehensive angiographical examination can enhance overall prognosis in presurgical planning in association with other vascular abnormalities.

Keywords: Anatomy, arterial fenestration, nonaneurysmal, vascular

INTRODUCTION

Anatomical variants of the cerebral arteries are encountered in a regular basis as an incidental finding.^[1] Fenestration means a vascular variation that begins with a common origin and then splits into two parallel luminal channels and re-joins distally.^[2] Each channel has its endothelium and tunica media but may share the adventitia as well. These two channels reunite distinctly. The etiology is thought to be due to the failure of midline fusion during embryogenesis, but this is controversial, depending on the site of fenestration.^[1] It differs from duplication that the latter variation also has a duplicated origin^[2] [Table 1].

The association of fenestrations with aneurysms has been extensively reported in the case of the series, although the exact relationship is not well-defined in studies with large samples.^[3-6] Probable association of fenestration with an aneurysm due to the presence of focal defects on the media layer at the proximal and distal end of the fenestrated segments is challenging for neurosurgeons and also interventional neuroradiologist.^[7]

Aim of the study

Focus the light on a rare entity of congenital vascular anomaly and its association with other anomalies or with cerebrovascular pathologies. The association between intracranial arterial fenestration (ICF) and the saccular aneurysm is frequently the focus of reports; we conduct a review of nonaneurysm conditions or anomalies that are reported to be associated with arterial fenestration demonstrating its clinical significance.

METHODS

A PubMed search of the English literature was conducted for 1970–2017 publications describing the cases of ICF and any

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Table 1: Classification of normal variants of the cerebral arteries

Normal variants	Details
Variation in vessel origin	Common origin Infundibulum Anomalous origin due to persistent embryological circulation
Variation in the number of vessels	Reduced number of vessels Increased number of vessels
Variation in morphology	Hypoplasia Hyperplasia Early bifurcation Fenestration
Variation in pathway	Persistent carotid vertebrobasilar anastomosis Persistent internal-external carotid anastomosis

associated condition or other vascular anomalies associated with it. The analysis excluded all the articles and case reports describing ICF and associated with intracranial aneurysm. We also excluded the reports of fenestrations without associated anomalies or pathologies. Data were collected from published cases on patient presentation, fenestration anatomic location, associated pathologies, and other vascular anomalies if present.

RESULTS AND DISCUSSION

We have conducted a comparative review, seeking the reported cases so far regarding the association of ICF with nonaneurysmal anomalies or pathology^[1,8-16] [Table 2].

The presence of vascular anomalies may imply an existing “segmental vulnerability,” which was explained by Lasjaunias. The segmental vulnerability is due to regional specificity in vascular anatomy and regional differences in responses to biological stimuli that affect the vascular environment. Such specificity generates an invisible discontinuity in the homogenous anatomical and histological hemodynamic system.^[17]

ICFs are a rare anatomic variant and remain indeterminate and likely heterogeneous clinical significance. They more often manifest in the anterior communication arterial complex and basilar artery (BA).^[18] Up to now, the frequency of fenestrations has been studied with cadavers^[19-21] and conventional angiographic series.^[22,23] The rate of fenestrations based on published conventional angiographic series has been much lower than that at cadaveric series.^[2]

Embryologically, the artery is plexiform until approximately the 18-mm stage, when it gradually becomes a single channel at the 21–24-mm stage e.g., anterior communicating artery (AcomA). Incomplete fusion of this plexus may result in either segmental or complete doubling of this artery. Fenestration of the BA is thought to be a result of incomplete fusion of the two primitive longitudinal arteries in the embryo at about the 5–9-mm stage.^[24] The vertebral

artery (VA) forms from an anastomosis between the cervical intersegmental arteries during the 7–12-mm stage, at which point there is a transient basilar-VA anastomosis. A remnant of this channel may explain such duplication. An anterior cerebral artery (ACA) fenestration may form because of the persistence of plexiform anastomosis between the primitive olfactory artery and the ACA. Whether this type of association represents concurrent congenital anomalies of embryological fusion or is purely coincidental remains a point of debate.^[15] Segmentally, unfused arteries constitute a lack of fusion of embryologically paired vessels. Duplications can occur where the “double-lumen” is due to two embryologically different vessels that fuse during development and an additional vessel persists. This is distinct from the fenestration where the two lumina correspond to a single artery.^[16]

The most common site of fenestration is the vertebrobasilar (VB) system, with the high prevalence reported in previous studies. Bharatha *et al.* reported a 2.8% overall rate of ICF.^[14] The issue of association of fenestrations with aneurysms has been found interesting by neuroradiologists and neurosurgeons for many years.^[2] Earlier studies have reported a high incidence of aneurysms in association with arterial fenestrations. Pathologically, it could be explained by the defects in the vessel wall at each end of the fenestration, which results in hemodynamic stress and structural degenerative changes in the vessel. It could be assumed that although fenestration is recognized as a risk for aneurysmal development, a significant association cannot be established^[25] [Table 3].

A fenestration is defined as a single artery with two luminal channels and may be due to a nerve or other anatomical structures “piercing” the artery.^[26] Theoretically, VA fenestration may mimic dissection. In both cases, there are two canals; however, in dissection, an intimal flap was usually too small for visualization as the second canal; this is confirmed by Teasdale *et al.*^[26] The actual prevalence of this anomaly is probably higher. The low reporting incidence is due to a lack of clinical implications themselves, examining only the upper part of the neck during routine cerebral angiography or the presence of tiny fenestrations called arterial splits, assumed to be intraluminal defects.^[11,27] Although very small fenestrations are called “arterial splits,” fenestrations-occupying long parts of the vessel and form two apparent independent arteries known as a big fenestration or “segmental duplications.” Even more picturesque names can be found in the literature such as a “buttonhole” or “island” formation, and the terms “bypass anomaly” and “diastematoarteria” have also been used. Recently, Vasović has proposed that the V1 segment fenestration is called a segmental duplication, and the V4 segment fenestration the “arteria vertebralis unfenestrated.” The names “pseudofenestration” or “extreme fenestration” of the VA are also used.^[28-31]

The complete occurrence of this well-known anatomy is not frequent, and it is important to know anatomical variations while interpreting imaging studies and before interventions.

Table 2: Characteristics of fenestrated intracranial arteries were associated with nonaneurysmal vascular pathology or anomalies reported in the literature between 1970 and 2017

Author(s)	Year	Location of fenestration	Associated anomaly or pathology	Remarks and comments
Mizukami <i>et al.</i> ^[8]	1972	VA	Cerebral arteriovenous malformation	There is an aneurysm in conjugation in the same patient
Yoshimoto <i>et al.</i> ^[9]	1992	VA	Enlarging cerebellar arteriovenous malformation	-
Yong and Seong ^[10]	2000	Left VA	Ruptured arteriovenous malformation at a right cerebellopontine angle	AVM supplied by the right AICA and PICA
Uchino <i>et al.</i> ^[11]	2001	VA VA VA + BA + left MCA	AVM	The frequency of coexistence of AVM and VA fenestration is 7%, 3 of 43
Okahara <i>et al.</i> ^[12]	2002	BA	Infarction (Wallenberg syndrome) Infarction at AICA territory Infarctions at ipsilateral posterior cerebral cortical artery Asymptomatic with persistent primitive trigeminal artery	
Chen <i>et al.</i> ^[13]	2007	Supraclinoid internal carotid artery	Ischemic stroke	Associated with ICA an aneurysm
Bharatha <i>et al.</i> ^[14]	2008	NS	AVM Dural AVF Azygous ACA	All of these were remote from the site of fenestration.
Miu ^[15] Other not-specified case reports ^[1,16]	2014	NS Persistent primitive hypoglossal artery P2 segment of fetal origin AICA origin displaced to the PICA–VA junction Hypoplastic VA Dolichoectatic BA VA originating from the aorta VA duplication Bilateral accessory MCA Hypoplastic A1 segment Premoyamoya-like changes Fused vertebrae	Venous angiomas	

VA: Vertebral artery, AVM: Arteriovenous malformation, AICA: Anterior inferior cerebellar artery, PICA: Posterior inferior cerebellar artery, BA: Basilar artery, MCA: Medial cerebral artery, ACA: Anterior cerebral artery, NS: Not specified, AVF: Arteriovenous fistula

Table 3: Significance of normal variants and anomalies^[1]

Normal variants and anomalies	Clinical significance
Anatomical pattern and differentiation from pathology	Anatomy knowledge and proximity of variants to other structures Variant versus pathology
The hemodynamic effect	Collateral function Hypoplastic arteries and stroke Obscure stroke incidents
Association with vascular and nonvascular disorders	Association with aneurysms Association with other vascular anomalies Association with congenital anomalies Association with other disorders
Preoperative planning for surgery and interventional procedures	Risk of disastrous hemorrhage Endovascular access and avoiding complications in interventional procedures

Few studies investigated the association between ICF and the vascular pathologies or anomalies other than an aneurysm

and its possible significance.^[32] The VA fenestration is often associated with other anomalies of the brain, spinal cord, and spine; these include fused vertebrae, other vascular anomalies, and an increased prevalence of aneurysms and vascular malformations.^[33] Uchino *et al.* have reported the frequency of coexistence of arteriovenous malformation and VA fenestration was 7%.^[11] Uchino *et al.* remark that atherosclerosis might hinder fenestration in the elderly.^[34] There has also been speculation about associations between VB system fenestrations and brainstem ischemia, or infarctions, although their relationships are still controversial.^[35] Tanaka *et al.* in a neuroradiological analysis of 23 BA fenestrations cases noted three cases of atherothrombotic infarctions (13%) in the territory of the VB system.^[36] The hemodynamic stress resulting from the fenestration of the feeding system of AVM might be an important factor in the enlargement of the small cerebellar AVM. An AVM associated with fenestration of the cranial arteries may more likely to bleed than AVM without this anomaly.^[10] The pathological (hyalinized) vessels in the small AVM probably succumbed to pressure within the vessels induced by the hemodynamic stress resulting from fenestration

of the VA, with consequent enlargement to a typical AVM.^[9] In an angiographic study of 51 patients, Uchino *et al.* reported a 7% prevalence of VA fenestration in cases of known arteriovenous malformations.^[11] Moreover, reports of VA fenestration have been associated with epidermoid cysts, other fenestrations, persistent trigeminal artery, neuralgia, VB ischemia, and agenesis of the corpus callosum.^[31]

The angiographic incidence of all fenestrations has been reported to be 0.3%–0.9%, whereas autopsy studies show the incidence of AcomA fenestrations to be 7.5%–40%, BA fenestrations 1%–5%, and VA fenestrations to be 1%–2%.^[13,24] Because of their low fenestration rates compared to cadaver series, and lack of data from nonaneurysmal patients, de Gast *et al.* could not establish a definite relationship.^[37] The higher sensitivity of postmortem analysis and the under-reporting of fenestrations on angiography can explain such discrepancies, as fenestrations are often asymptomatic unless accompanied by the asymptomatic aneurysm.^[38] On the other hand, the prospective identification of fenestrations, since they may result in complex anatomy, can be critical for planning surgical or endovascular procedures with safety.^[39]

CONCLUSION

Variations in the arterial system, whether normal or anomalies, are not uncommon angiographic findings, and these findings may be a warning sign for other associated more complex pathologies. A thorough and sophisticated preprocedural or presurgical study of the patient's angiography may increase the possibility of discovering nonaneurysmal-associated anomalies and then include these findings as intimate parts of management planning to improve the overall prognosis and minimize the possibility of unfavorable surprises. For sure, the firmer conclusion can certainly be drawn with continuing reports of such associations and their clinical significance.

Highlights

- Variations in the arterial system are common findings
- Vascular anomalies may translate into a segmental vulnerability
- Vertebral artery fenestration may mimic dissection
- Vascular variations potentially endanger the patient during the procedure
- Preprocedural angiography may be of benefit to avoid harm for patients.

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Conflicts of interest

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