Fenestration of intracranial arteries with special attention to associated aneurysms and other anomalies.

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Fenestration of Intracranial Arteries with Special Attention to Associated Aneurysms and Other Anomalies

William P. Sanders,1 Patrick A. Sorek,1 and Bharat A. Mehta1

PURPOSE: To determine the association of intracranial arterial fenestration and aneurysms.

METHODS: We retrospectively reviewed 5,190 cerebral angiogram reports and identified 37 patients with arterial fenestrations. RESULTS: These 37 patients had 38 fenestrated arteries: 16 basilar, 10 vertebral, nine middle cerebral, and three anterior cerebral arteries. Seven of these patients had a total of 13 aneurysms, although only one aneurysm was at the site of a fenestration. The remaining aneurysm patients had fenestrations as unassociated findings. Other anomalies detected were two azygous anterior cerebral arteries, one dural arteriovenous shunt of the cavernous sinus, one extracranial arteriovenous fistula, and one developmental venous anomaly (venous angioma). None of the fenestrations were in the vessels directly involved with these associated lesions. The incidence of aneurysm at the fenestration was 7% (one in 16) for basilar artery fenestrations. Considering all fenestrations, the incidence of aneurysm at the site of fenestration was 3%. CONCLUSION: Our data show that the association of a fenestration with an aneurysm at the fenestration site is not different from the typical association of circle of Willis bifurcations with saccular aneurysms.

Index terms: Aneurysm, cerebral; Arteries, abnormalities and anomalies; Arteries, anatomy; Arteries, cerebral

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Fenestration of a cerebral artery refers to a division of the lumen of an artery, resulting in two distinct endothelium-lined channels, which may or may not share their adventitial layer. There is a spectrum of appearances, from a tiny island of tissue separating the two channels to actual duplication of a long segment of the involved artery. The embryologic basis of fenestration of the basilar and vertebral arteries has been elucidated by Padget (1), but the etiology is less clear for fenestration of the middle and anterior cerebral arteries (MCA and ACA). There is evidence in the literature suggesting a relationship of fenestration to intracerebral aneurysm (2–11). We report 38 fenestrations of intracerebral vessels and discuss associated lesions encountered in cases drawn from a 5½-year period.

Methods

Retrospective review of the reports of 5,190 cerebral angiograms performed over a 5½-year period yielded 37 patients with 38 arterial fenestrations or duplications. These 37 angiograms were then carefully reviewed for the presence and location of fenestrations, aneurysms, and other developmental variants.

Results

The 38 fenestrations were found in 37 patients 18 to 77 years old, 21 men and 16 women. The locations of the fenestrated vessels are shown in Table 1. There were seven patients with a total of 13 aneurysms; there was only one aneurysm at the site of a fenestration, in the midbasilar artery (Fig. 1). The remainder of the aneurysms were clearly separate from the fenestrations. In fact, only two of these aneurysms were in the same arterial tree as the fenestrations: one basilar tip aneurysm with a fenestration proximal to the superior cerebellar arteries and another basilar tip aneurysm with fenestration at the level of the anterior inferior cerebellar arteries. The incidence of aneurysm in our 16 basilar artery fenestration
sites was one in 16 (7%). Considering all intracranial fenestrations, the incidence of aneurysm at the fenestration site was one in 38 (3%).

Other associated anomalies included two azygous anterior cerebral arteries, one extracranial vertebral arteriovenous fistula, one dural arteriovenous shunt of the cavernous sinus, and one developmental venous anomaly (venous angioma). None of these anomalies were in the same vascular tree as the fenestrated artery. No history of trauma was present in the patients with the dural arteriovenous shunt or the vertebral arteriovenous fistula, and these lesions are presumed to be developmental in etiology.

Cerebral angiographic studies were performed for a variety of reasons, including 17 for ischemic symptoms, seven for subarachnoid hemorrhage, five for headaches without hemorrhage to rule out aneurysm, and the rest for nonspecific symptomatology in which a vascular malformation or neoplasm were considerations.

Discussion

Basilar Artery Fenestration

The basilar artery is formed by fusion of the bilateral longitudinal neural arteries (Fig. 2A) during approximately the fifth week of fetal growth (1). During this fusion process there are temporary bridging arteries connecting the longitudinal neural arteries that regress as fusion is completed (Fig. 2B). Fenestrations occur at any segment along the basilar artery in which there is failure of complete fusion of the medial aspects of the longitudinal neural arteries. Postmortem exami-

TABLE I: Location of arterial fenestrations

<table>
<thead>
<tr>
<th>Artery</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Basilar</td>
<td>16</td>
</tr>
<tr>
<td>Proximal</td>
<td>5</td>
</tr>
<tr>
<td>Mid</td>
<td>7</td>
</tr>
<tr>
<td>Distal</td>
<td>4</td>
</tr>
<tr>
<td>Vertebral</td>
<td>10</td>
</tr>
<tr>
<td>Middle cerebral</td>
<td>9</td>
</tr>
<tr>
<td>Anterior cerebral</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td>38</td>
</tr>
</tbody>
</table>

Fig. 1. Aneurysm associated with basilar artery fenestration, right vertebral artery injection.

A, Frontal view. Fenestration is clearly seen (arrowhead) with aneurysm (arrows) superimposed over basilar artery.

B, Lateral view. Anteriorly projecting aneurysm is readily appreciated (arrow).
FENESTRATION OF INTRACRANIAL ARTERIES

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Bridging a.
Longitudinal neural a.
(Basilar a. in formation)
Otic vesicle
Int.carotid a.
hypoglossal a.
Paired aorta

12.5mm Embryo

Ant.cerebral a.
Middle cerebral a.
Basilar a.

14mm Embryo

Vertebral a.
Ant.spinal a.

Fig. 2. A and B, Embryologic development of the basilar artery (reprinted with permission from Padget (1)). Frontal schemata demonstrate the temporary bridging arteries that regress as the bilateral longitudinal neural arteries fuse to form the basilar artery during the fifth week of fetal growth.

nation of these fenestration sites has revealed normal architecture of the lateral walls of the basilar artery with a medial septation blending into two distinct channels that may or may not be of equal size (Fig. 3) (10, 12, 13). Histologic examination has demonstrated normal layers in the medial arterial walls, except for a short length at the proximal and distal ends of the central septation, in which there are defects in the media similar to bifurcations of normal cerebral arteries (10). The dual channels may or may not share a common adventitia depending on the degree of separation of the two limbs of the fenestration (10, 12).

One postmortem study demonstrated that almost 6% of 220 cases had fenestration of the basilar artery (14). However, the angiographic incidence of basilar fenestration has been reported to be from 0.04% to 0.6% (15, 16); our incidence was 0.3%. The discrepancy between postmortem and angiographic incidence is likely related to the increased sensitivity of postmortem examination. In addition, a review of angiographic reports to identify fenestrations would also likely lead to a lower angiographic incidence because of underreporting of fenestrations on the original study.

There are multiple case reports describing aneurysms associated with basilar artery fenestration (2–11). The most impressive data was presented by Campos et al in which 21 of 59 aneurysms at the vertebrobasilar junction were at the proximal end of a fenestration (2). These are rare aneurysms that are not located at typical bifurcations or sites of origin of other basilar artery branches. In their series, patients with known aneurysms were evaluated for the pres-

Fig. 3. Left vertebral artery injection, frontal view. Unequal size of the fenestration channels (arrows) in the proximal basilar artery.
ence of fenestration. In our series, patients with fenestrations were evaluated for the presence of aneurysm. Campos’s series is compelling evidence that when these vertebrobasilar junction aneurysms are present, a fenestration will be commonly identified. However, their observations were biased in that all of their patients were referred with known aneurysms whereas our patients were selected according to the presence of fenestrations. None of the five proximal fenestrations in our series were associated with aneurysm. Perhaps a series with a greater number of basilar artery fenestrations would reveal more aneurysms.

We did encounter an anteriorly directed aneurysm associated with a midbasilar fenestration (Fig. 1). This aneurysm is similar in appearance to those reported by Campos et al, although slightly more distal in the basilar artery than their series. The overall incidence of aneurysm associated with basilar fenestration was 7% in our series.

Most authors postulate that there is a predisposition to aneurysm formation at fenestrations for two reasons: a defect in the media of the proximal and distal ends of the fenestrated segment, and turbulent flow at the “bifurcation” created by the proximal end of the fenestration (2, 5, 10, 12, 13, 15). Indeed, these are the same accepted mechanisms for saccular aneurysm formation at bifurcations of intracranial arteries. Perhaps a fenestration should be considered as an additional, albeit rare, bifurcation in regard to aneurysm formation. An interesting report described a fenestration at the vertebrobasilar junction not associated with an aneurysm, with the appearance suggestive of persistence of a primitive lateral vertebrobasilar anastomosis (17). The lesion shown in that paper had more of the appearance of what we describe as vertebral artery duplications, rather than basilar artery fenestrations.

Vertebral Artery Duplications

The vertebral arteries are formed between the fetal ages of 32 and 40 days, by interconnecting several primitive cervical segmental arteries. In the 5- to 6-mm embryo these vessels provide the proximal supply to the longitudinal neural arteries via the first cervical segmental arteries and the transient basivertebral anastomotic vessels (Fig. 4A) (1, 3). Later, as more cervical segmental arteries develop, anastomoses are formed connecting the cervical segmental arteries to form the primitive vertebral arteries (Fig. 4B).

The segmental origin of the primitive vertebral arteries gives rise to the possibility of duplication on the basis of failure to involute the transient vessels near the skull base, especially the basivertebral anastomotic vessels (3, 17). The extracranial duplications are most likely caused by persistence of cervical segmental arteries whereas the intracranial components probably arise from persistent basivertebral anastomoses.

All of our 10 vertebral fenestrations were at the C1 level or above (Fig. 5), similar to other reports (3, 18). The reasons for predisposition to distal segment duplication are unknown at this time. These duplications have not yet been histologically scrutinized to our knowledge. No aneurysms were associated with our 10 vertebral artery fenestrations.
Anterior and Middle Cerebral Artery Fenestration

The distal primitive internal carotid artery divides into several branches, the largest being the anterior choroidal artery, followed by multiple twigs representing the primitive middle and anterior cerebral arteries (1). The small transient branches disappear, leaving single medial and lateral divisions of the internal carotid artery. Although there is not a proven association, it is probable that persistence of more than one of the primitive channels results in the presence of fe-
nestrations and duplications of the proximal segments of the middle cerebral artery and anterior cerebral artery.

There were 14 cases of anterior cerebral artery fenestrations (Fig. 6) and one case of middle cerebral artery fenestration (Fig. 7) reported in a series of 220 autopsy specimens, for an incidence of 7% (14). In a previous series of 4,500 angiograms reported by Teal et al, one middle cerebral artery and two anterior cerebral artery fenestrations were reported for an incidence of 0.07% (15). The incidence of anterior circulation fenestration in our series was 0.2%. No aneurysms were associated with the 12 anterior fenestration sites in our patients.

The angiographic distribution of fenestrations of the intracranial arteries in our study parallels the distribution based on autopsy data. Variations in incidence are likely technical. Aneurysms may be associated with fenestrations just as they are associated with the usual arterial bifurcations as the histologic similarity has been demonstrated (10). Our data show that the association of a fenestration with an aneurysm at the fenestration site is not different from the typical association of vascular bifurcations with saccular aneurysms.

References