Non-Atherosclerotic Fusiform Cerebral Aneurysms

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ABSTRACT: Background: Fusiform cerebral aneurysms are dilatations of the entire circumference of a segment of cerebral artery, usually considered due to atherosclerosis in adults. They are relatively thick-walled and elongated, causing neural compression or ischemia when discovered. We have noted a subset of fusiform cerebral aneurysms that vary from this common description. Patients: Out of a series of 472 intracranial aneurysms treated over 11 years, 11 patients between the ages 16 and 67 years (mean age 37) were identified who had discrete fusiform aneurysms unassociated with generalized cerebral atherosclerosis, connective tissue disorder or inflammation. Three presented with hemorrhage, six with neural compression by the aneurysm and two were discovered incidentally. Results: Nine aneurysms were located in the posterior circulation, the other two in the intracranial carotid artery. Their mean length and width were 16.3 and 11 mm, respectively. Three aneurysms contained thrombus. The eight aneurysms that were exposed surgically were partly or substantially thin-walled with normal appearing parent arteries. Eight were treated with proximal occlusion and three were circumferentially “wrapped”. Parent artery occlusion caused one death and one mild disability and the remaining patients made good recoveries (follow-up 0.5 - 10 years). Conclusions: There is a subset of cerebral aneurysms with discrete fusiform morphology, apparently unrelated to cerebral atherosclerosis or systemic connective tissue disease, thin-walled in part or whole, more common in the vertebrobasilar system, and possessing a risk of rupture. Treatments currently available include proximal occlusion or aneurysm “wrapping”, different approaches than neck-clipping or endovascular coiling of side-wall saccular cerebral aneurysms that leave the parent artery intact.

any other systemic and/or heritable connective tissue or vascular disease. Like atherosclerotic dolichoectasias, these aneurysms are commonest in the vertebrobasilar circulation and frequently cause compressive symptoms but, unlike them, these aneurysms occur in younger patients, are relatively discrete and arise from otherwise normal arteries. The series of patients presented here suggests that not all aneurysms with fusiform morphology have a clear atherosclerotic etiology or are related to a detectable connective tissue disease.

**PATIENTS**

Out of a series of 472 intracranial aneurysms treated by a single neurovascular surgeon over 11 years, and not including six additional patients with intracranial arterial dissections, 11 patients (2.3% of the total) between the ages of 16 and 67 (mean age 37) were identified who had discrete fusiform intracranial aneurysms which were unassociated with detectable cerebral atherosclerosis or a diagnosed hereditary or acquired connective tissue or inflammatory disorder, such as Marfan’s syndrome, pseudoxanthoma elasticum, Ehlers-Danlos syndrome, neurofibromatosis, or any type of intracranial infectious or inflammatory condition. Specifically, cerebral atherosclerosis was considered very unlikely when the aneurysm appeared discrete and there was no evidence of either diffuse cerebrovascular disease or atherosclerosis elsewhere as determined by cerebral arterial imaging and/or by direct

![Figure 1](https://example.com/figure1.jpg)
Table: Summary of Patient Series

<table>
<thead>
<tr>
<th>Patient name, age</th>
<th>Clinical Presentation</th>
<th>Aneurysm location, length &amp; width (mm)</th>
<th>Intraluminal thrombus</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Mrs. L.K., 67 y.o.</td>
<td>SAH</td>
<td>left PCA, P1 and P2 segments 8 x 3.5 mm</td>
<td>no</td>
<td>saccular bleb clipped, remainder wrapped</td>
<td>Good, 10 years f/u</td>
</tr>
<tr>
<td>2. Mr. A.B., 16 y.o.</td>
<td>Painful right 3 &amp; 5 cranial neuropathies</td>
<td>right cavernous ICA, 20 x 11 mm</td>
<td>no</td>
<td>EC-IC bypass, proximal ICA occlusion</td>
<td>Good, 6 years f/u</td>
</tr>
<tr>
<td>3. Mrs. S.T., 47 y.o.</td>
<td>Spastic left hemiparesis, tremor</td>
<td>right PCA, P1 and P2 segments, 30 x 22 mm</td>
<td>yes</td>
<td>proximal PCA clip occlusion</td>
<td>Good, 5 years f/u</td>
</tr>
<tr>
<td>4. Mr. V.M., 38 y.o.</td>
<td>left 9, 10 &amp; 11 cranial neuropathies, facial numbness, vertigo</td>
<td>left intradural VA, 12 x 4 mm</td>
<td>no</td>
<td>wrapped</td>
<td>Good, 5 years f/u</td>
</tr>
<tr>
<td>5. Mrs. D.G., 18 y.o.</td>
<td>left 3 &amp; 5 cranial neuropathies</td>
<td>left SCA, 10 x 6 mm</td>
<td>yes</td>
<td>endovascular coil occlusion (x 2)</td>
<td>Good, 3 years f/u</td>
</tr>
<tr>
<td>6. Rev. E.E., 49 y.o.</td>
<td>incidental</td>
<td>right intradural VA, 10 x 5 mm</td>
<td>no</td>
<td>proximal clip occlusion</td>
<td>Good, 2 years f/u</td>
</tr>
<tr>
<td>7. Mrs. L.G., 47 y.o.</td>
<td>recurrent SAH</td>
<td>left PCA, P2 segment, 3 x 5 mm</td>
<td>no</td>
<td>proximal and distal clip occlusion</td>
<td>Good, 2 years f/u</td>
</tr>
<tr>
<td>8. Mrs. A.S., 46 y.o.</td>
<td>right-sided weakness</td>
<td>left PCA, P2 segments, 30 x 20 mm</td>
<td>yes</td>
<td>proximal clip occlusion</td>
<td>Good, 20 months f/u</td>
</tr>
<tr>
<td>9. Mr. G.B., 24 y.o.</td>
<td>recurrent SAH</td>
<td>BA trunk, 30 x 25 mm</td>
<td>no</td>
<td>proximal clip occlusion</td>
<td>Died</td>
</tr>
<tr>
<td>10. Mr. D.V., 53 y.o.</td>
<td>right hemiparesis</td>
<td>VA-BA junction, 19 x 15 mm (both terminal VAs)</td>
<td>no</td>
<td>right VA balloon occlusion</td>
<td>Good, 1 year f/u</td>
</tr>
<tr>
<td>11. Mrs. M.J., 43 y.o.</td>
<td>incidental</td>
<td>left terminal ICA, 7 x 4 mm</td>
<td>no</td>
<td>wrapped</td>
<td>Good, 6 months f/u</td>
</tr>
</tbody>
</table>

SAH = subarachnoid hemorrhage, ICA = internal carotid artery, VA = vertebral artery, BA = basilar artery, PCA = posterior cerebral artery, SCA = superior cerebellar artery, P1 = precommunicating segment of the posterior cerebral artery, P2 = post-communicating segment of the posterior cerebral artery to the quadrigeminal cistern.

*Glasgow Outcome Scale*39

inspection of the parent and adjacent arteries at the time of surgery. At the time of clinical presentation and in follow-up, connective tissue disorders were ruled out on the basis of standard clinical criteria.8 None of the patients had detectable intracranial sepsis.

Three patients presented with subarachnoid hemorrhage, six with symptoms and signs of either cranial nerve or brain stem compression by the aneurysm and two aneurysms were incidental discoveries (Table). None had calcification of the aneurysm wall and three had intra-aneurysmal thrombus (laminated clot) prior to treatment (Figure 1). Seven of the 11 patients underwent preoperative magnetic resonance imaging (MRI) (patients no.s 2, 3, 4, 6, 8, 10 and 11) and none had evidence of cerebral infarction related to the aneurysm. Angiographic assessment of every patient and direct surgical inspection of the eight patients who underwent surgery indicated normal parent arteries and normal arteries adjacent to the fusiform aneurysm. The mean length and width of the aneurysms was 16.3 mm and 11 mm, respectively. In recent years we have found three-dimensional computerized tomographic angiography (CTA) very useful in the anatomical characterization of these aneurysms.
TREATMENT AND OUTCOME

All of the patients in this series were treated in some manner. Eight patients underwent proximal parent artery occlusion, five with microsurgical clipping (one of whom also underwent trapping of a small fusiform aneurysm [patient no. 7]), two with proximal balloon occlusion and one with intra-aneurysmal endovascular Guglielmi detachable (Target Therapeutics, Fremont, CA) coil embolization (Figure 2). Three aneurysms were circumferentially wrapped with cotton sheets that were held clipped or stapled around the fusiform aneurysm (Figure 3). One of these patients (patient no.1) also had a saccular bleb arising from her posterior cerebral artery fusiform aneurysm (thought to have caused her subarachnoid hemorrhage) that was clipped directly prior to wrapping the entire fusiform segment of artery. One patient, no. 2, underwent a superficial temporal artery-middle cerebral artery bypass prior to balloon occlusion of a cavernous segment internal carotid artery aneurysm.

Outcome has been generally good. One patient with a giant basilar trunk aneurysm who underwent proximal basilar artery coil occlusion suffered brain stem infarction and died several days following surgery (patient no. 9) (Figure 4). Another with a large...
“pantaloon” aneurysm of the vertebrobasilar junction suffered a small medullary infarction following occlusion of the right vertebral artery, from which he made a good recovery (patient no. 10) (Figure 5). Follow-up between six months and ten years has not detected clinical aneurysm recurrence in any patient.

**DISCUSSION**

Intracranial arterial aneurysms are classified and described both etiologically and morphologically and, for many aneurysms, there is a correlation between their cause and shape. The most common type of intracranial aneurysm, making up over 90% of the total, are “developmental” in origin, acquired rather than congenital lesions arising at arterial bifurcations, branch points or curves. These locations are subjected to particular hemodynamic stresses that over time lead to focal vessel wall degeneration and aneurysm formation.9,10 Developmental aneurysms have a saccular morphology (hence the older term “berry” aneurysms), can be of any size, and have an orifice and neck arising from an underlying normal parent vessel.

Fusiform intracranial aneurysms have been characterized as thickened, widened and elongated arteries due to atherosclerosis in an older population (geriatric men most commonly), occurring most often in the vertebrobasilar arteries where they can become quite large and compress cranial nerves or brain stem causing symptoms such as trigeminal neuralgia, hemifacial spasm, deafness, or sensorimotor dysfunction of the pharynx, larynx or limbs.4,11-14 Intraluminal thrombosis and embolism can cause ischemia and infarction.4,5 Histopathological examination of the walls of these aneurysms shows deficiencies in the elastic lamina, fibrotic replacement of the muscularis, calcium and lipid deposition, mural hemorrhage and infiltration with both acute and chronic inflammatory cells.6,15-17 Although quite uncommon, cerebral arterial ectasia and fusiform intracranial aneurysms have also been reported to occur in younger persons with certain systemic connective tissue disorder, including Ehlers-Danlos disease.
syndrome, Marfan’s syndrome, pseudoxanthoma elasticum, tuberous sclerosis and neurofibromatosis.\textsuperscript{18,19}

Intracranial arterial dissections can result in stenosis, occlusion, double-lumens and also fusiform arterial dilatations causing subarachnoid hemorrhage when the adventitial layer is breached. The possibility that some of the aneurysms described in this report were intracranial dissecting aneurysms must be considered, especially since intracranial dissections more commonly occur in the vertebral and basilar arteries.\textsuperscript{20-22} The majority of patients with intracranial arterial dissections (and all

\textbf{Figure 4:} A 24-year-old healthy young man (patient no. 9) collapsed while motor-bike racing and, within 24 hours, suffered a recurrent subarachnoid hemorrhage from a large mid-basilar artery aneurysm (upper left panel, left vertebral angiogram). At surgery it was found that the aneurysm was fusiform so the proximal basilar artery was clip-ocluded, since it was known that the patient had posterior communicating arteries to provide collateral flow to the upper brain stem. The aneurysm thrombosed following, causing brain stem infarction and the patient died several days later. Autopsy confirmed the fusiform nature of the aneurysm (upper right, showing the opened aneurysm, clips occluding the proximal basilar artery and a wooden stick [arrow] inserted into the distal basilar artery lumen). Pathological examination (lower panel, Verhoeff Van Gieson staining) showed the proximal parent basilar artery (A) with normal tunica intima (i), elastic lamina (arrow head), tunica media of smooth muscle (m) and tunica adventitia (a). The aneurysm neck begins to lose elastic lamina and tunica media (B), and the aneurysm wall shows a complete loss of these two elements (C), replaced by degenerative fibrous tissue. No changes consistent with atherosclerosis were found.
of the 65 patients with typical dissections in Mizutani et al’s report21) present with subarachnoid hemorrhage and imaging demonstrates irregular luminal stenosis at either end of a fusiform enlargement, often with a tell-tale intimal flap. Among the three patients in our series who presented with subarachnoid hemorrhage, one died and pathological examination of the basilar aneurysm ruled out a dissection, and the radiological appearances of the other two posterior cerebral artery aneurysms that ruptured were not typical of dissection (as described above). Nevertheless, in the absence of neuropathological proof, it remains possible that these and perhaps other fusiform aneurysms in our series were atypical intracranial arterial dissections.

There are a number of other more rare and special intracranial aneurysms, including traumatic,23 neoplastic,24,25 inflammatory,26,27 infectious28-31 and radiation induced types,32 where focal arterial wall injury, invasion or destruction leads to aneurysmal dilatation that is usually irregular and sometimes fusiform in morphology. However, these types of aneurysms are quite unlike the fusiform aneurysms described in this report, arising usually in peripheral arteries, normally only several millimeters in size and, because of their distal location, causing parenchymal hemorrhage when they rupture.

The patients presented in this report are unique in that their aneurysms had distinct fusiform morphologies but there was no evidence of adjacent or widespread cerebral atherosclerosis, nor was there any clinical evidence of connective tissue disorder, localized infection or vessel wall inflammation. Also inconsistent with an atherosclerotic etiology was the young mean age of our patients (37 years) as well as the absence of clinical or radiological evidence of cerebral ischemia associated with the aneurysms. It needs to be acknowledged, however, that without histopathological proof obtained at autopsy (available for only one patient in our series) we cannot absolutely rule out either atherosclerosis or occult systemic connective tissue disease. Long-term follow up of several patients suggests that once treated, the underlying process does not spread to adjacent vessels. It is interesting that the aneurysms in this series showed a proclivity for the posterior circulation, and the posterior cerebral artery in particular, which is similar to the more common atherosclerotic fusiform aneurysms. Similar findings to these were recently reported by Drake and Peerless.33 Although the 120 fusiform intracranial aneurysms they treated surgically were all giant (≥ 2 cm) in size, they found only three due to an identifiable ateriopathy and only six in patients with atherosclerosis. In the remaining 111 patients the etiology of their giant fusiform aneurysms could not be determined. In these much younger patients, with an almost even sex ratio, aneurysms predominated in the basilar trunk and posterior cerebral artery (90% affected the posterior circulation), and 20% bled, findings similar to our own series of smaller aneurysms. Idiopathic fusiform aneurysms affecting the middle cerebral artery were also described by those authors and have been reported by others,34-36 although we have not yet encountered one.

The morphology of fusiform aneurysms requires that treatment, when indicated by either the presence of symptoms (nine of the 11 patients in this series), or the predicted long-term risk of growth and possible rupture (two of the 11 patients in this series) consist of either proximal occlusion, proximal and distal occlusion (“trapping”), or circumferential wrapping with or without clip reinforcement to stabilize and strengthen the aneurysm wall.37,38 When Hunterian proximal occlusion or trapping are being considered, success is dependent upon adequate collateral circulation to both the distal vessels as well
as any branches or perforator vessels arising from the fusiform aneurysm itself. That collateral circulation may arise from the circle of Willis, bypass vessels created surgically, or natural leptomeningeal or deep arteriolar collateral flow. Treatment must be individualized to the patient, but basilar trunk aneurysms present the greatest difficulties while posterior cerebral artery fusiform aneurysms are often simply and effectively managed by proximal posterior cerebral artery occlusion, since this artery usually has good collateral blood supply.

The underlying cause of the type of fusiform aneurysms described in this report is unknown at this time but appears to bear no clear relationship to cerebral atherosclerosis. The principle aim of this report is to emphasize that fusiform and atherosclerotic aneurysms are terms that should not be used interchangeably. Fusiform atherosclerotic aneurysms should be distinguished from those that are fusiform in shape but idiopathic in etiology.

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REFERENCES