Giant serpentine aneurysm

Report of two cases

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The authors describe two cases of giant middle cerebral artery aneurysms presenting as mass lesions. Angiograms in each case revealed a distinctive serpentine vascular channel surrounded by an avascular area causing a “mass effect.” Both lesions were resected in toto with excellent clinical results. Similar lesions in the literature are noted and the pathophysiology and origin of this group of aneurysms are discussed.

KEY WORDS • giant aneurysm • serpentine channel • angiography • middle cerebral aneurysms

Contemporary literature on cerebral aneurysms tends to categorize a group of lesions identified as “giant aneurysms” on the basis of size alone (greater than 2.5 cm in diameter). However, within this group, a subgroup exists consisting of large partially thrombosed aneurysms with a persistent serpentine vascular channel. Case reports of lesions within this subgroup have been scattered throughout the recent literature.3,7,10 This paper attempts to clearly identify the subgroup on clinical and pathological grounds and to report successful total excision of two of these lesions.

Case Reports

Case 1

This 39-year-old man entered the hospital with a 2-week history of severe frontal headache accompanied by vertigo and fuzziness of vision. The past history was unremarkable. On examination he was normotensive, well developed, and in no acute distress. Bilateral papilledema was present; cranial nerves, and motor and sensory systems showed no deficits.

Plain x-ray films of the skull revealed a small right frontal calcification. An angiogram demonstrated opacification of the serpentine vascular channel in the Sylvian region (Fig. 1). A right frontotemporal craniotomy was performed and a large partially thrombosed aneurysm of the right middle cerebral artery was resected in toto. Postoperatively the patient exhibited a left hemiparesis and a partial left homonymous hemianopia. Two weeks later he was discharged and no neurological deficits were noted other than a partial left homonymous visual field cut. Ten years postoperatively, he was neurologically intact except for the same visual defect.

Case 2

This 30-year-old man was admitted to the hospital for “depression.” There had been a 6-month history of frontal and vertex head-
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FIG. 1. Case 1. Right carotid angiogram, anteroposterior (left) and lateral (right) views. Opacification of the serpentine vascular channel in the Sylvian region can be seen. The channel appears to arise from a branch of the middle cerebral artery; there is a large avascular region adjacent to the channel.

aches, but 5 days before admission the headaches became more severe and he experienced bouts of crying and difficulty in speaking. On examination he was alert and normotensive, with red puffy eyes, in an obvious state of anxiety. He was oriented to person, place, and time, but showed evidence of components of both receptive and expressive aphasia. He could not think of the names of friends or common objects and his attention span was short. A mild right central facial palsy and right hemiparesis were present.

Plain x-ray films of the skull showed a calcified pineal gland with a large shift from left to right. A brain scan showed a large area of increased activity in the left temporal area (Fig. 2). An angiogram showed opacification of the serpentine vascular channel (Fig. 3). At surgery a large aneurysmal mass was seen to occupy the area of the left Sylvian fissure. The parent feeding vessel was a branch of the middle cerebral artery at the trifurcation. Distally, the mass was associated with the angular branch of the middle cerebral artery. The feeding vessel was clipped and the mass was removed in toto. Immediately postoperatively the patient had a dense right central facial palsy and right hemiparesis, but within several hours, these deficits cleared. He was discharged 1 week later with only a mild diplopia. Postoperative angiography showed complete extirpation of the aneurysm.

Observations

Pathological Study

The specimens from both cases were very similar. The first measured $3.8 \times 3.6 \times 2$ cm.
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Fig. 3. Case 2. Left carotid angiogram. Left: Anteroposterior view shows opacified serpentine channel, square shift of anterior cerebral artery, stretching of insular vessels, and elevation of Sylvian point. The genu of the middle cerebral artery is deviated medially. Right: Lateral view is a later arterial phase demonstrating delayed flow through the vascular channel that ends in the angular branch of the middle cerebral artery.

and the second measured $6.5 \times 6 \times 5$ cm. Both were described as large globoid masses with walls measuring from 0.1 to 0.3 cm in thickness (Fig. 4 left). On sectioning, each mass contained an irregular serpentine channel coursing along and through the greater portion of the partially thrombosed aneurysm (Fig. 4 right). There were multiple small lumina noted in addition to the larger persistent channel in the surgical specimen.

Fig. 4. Case 2. Left: Intact gross specimen showing large, pear-shaped configuration of the mass. Small vessels can be seen on its outer surface. Right: Sections of surgical specimen showing that the mass is largely composed of old laminated thrombus. Note variation in thickness of the specimen wall and the large channel coursing on the outer surface of the mass (lower right). This well formed arterial channel communicated through a defect in its wall with the internal vascular channel of the large thrombosed aneurysm. A large portion of old thrombus is missing from the mass shown lower right.
TABLE 1

<table>
<thead>
<tr>
<th>Author</th>
<th>Age (yrs), Sex</th>
<th>Symptoms</th>
<th>Signs</th>
<th>EEG</th>
<th>Aneurysm Site</th>
<th>Follow-up Condition</th>
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<tr>
<td>Sadik, et al.,</td>
<td>47 M</td>
<td>dizziness, nausea, fatigue, depression, visual loss, somnolence; lt hemiparesis</td>
<td>decreased visual acuity; mild lt hemiparesis</td>
<td>slow bilaterally, most prominent in rt frontal</td>
<td>middle cerebral artery, rt</td>
<td>died 7th postop day</td>
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<tr>
<td>1965</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Cantu &amp; LeMay,</td>
<td>58 M</td>
<td>headache</td>
<td>dysnomia, dyscalculia, dyspraxia, finger agnosia</td>
<td>lt F-P slowing</td>
<td>middle cerebral artery, lt</td>
<td>nominal dysphasia</td>
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<td>1966</td>
<td></td>
<td></td>
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<tr>
<td>Terao &amp; Muraoka</td>
<td>38 M</td>
<td>headache, nausea &amp; vomiting, dysphasia; twitching lt arm &amp; leg</td>
<td>somnolence and lt hemiparesis</td>
<td>rt-sided slowing</td>
<td>middle cerebral artery, rt</td>
<td>died 6th postop day</td>
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<td>1972</td>
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<td>Lukin &amp; Chambers</td>
<td>67 M</td>
<td>headache &amp; depression</td>
<td>expressive dysphasia and rt hemiparesis</td>
<td>lt temporal slowing</td>
<td>middle cerebral artery, lt</td>
<td>rt hemiparesis &amp; expressive aphasia</td>
</tr>
<tr>
<td>1975</td>
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<tr>
<td>Segal &amp; McLaurin</td>
<td>39 M</td>
<td>headache, vertigo, fuzzy vision; numb lt arm &amp; leg</td>
<td>bilateral papilledema</td>
<td></td>
<td>middle cerebral artery, rt</td>
<td>neurologically intact except for partial field cut</td>
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<tr>
<td>1977</td>
<td></td>
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<tr>
<td></td>
<td>30 M</td>
<td>headache, depression, speech impairment</td>
<td>expressive &amp; anomic dysphasia; rt hemiparesis</td>
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<td>middle cerebral artery, lt</td>
<td>neurologically intact</td>
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from Case 1. These lumina appeared to be attempts at recanalization through the thrombosed portions. Sections through the larger clotted portions of each mass revealed evidence of old thrombosis within the laminated clot. The specimen from Case 2 showed arterial vessels coursing in the adventitia of the aneurysm apparently supplying the outer surface of the mass. Neither mass showed evidence of past or present rupture. There was no evidence of hematoma either surrounding or incorporated into the walls of either mass. Both aneurysms and the vessels coursing in their adventitia showed evidence of atherosclerosis.

The walls of the specimens consisted of thickened fibrous tissue, which was acellular for the most part. There was no internal elastic lamina, nor were there any remnants of an endothelial lining; however, several regions appeared to show evidence of cellular muscular elements within the aneurysm wall. Neither aneurysm arose from the typical apical medial gap found at arterial forkings. In this respect they differ from the most common variety of saccular aneurysms. It is interesting, however, that the largest but less frequent variety of saccular aneurysms do not arise from arterial forkings. They are usually related to severe generalized atherosclerosis in the cerebral vasculature and are often multiple. Furthermore, they are typically found in an elderly age group and involve the vertebral, basilar, and internal carotid arteries. Rupture is rare within this group.9

The lesions in our two patients (and those listed in Table 1) appear to differ from the saccular varieties in that they did not occur at arterial forkings, were not associated with generalized atherosclerotic changes, did not
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occur mainly in elderly age groups, and were concentrated in the middle cerebral artery distribution.

Radiological Aspects

The radiological picture was quite characteristic. A serpentine vascular channel was fed by the middle cerebral artery and ultimately ended in a small, but often important distal branch. There was slowed circulation through the vascular channel, but no evidence of vasospasm was seen.

A "mass effect" was nearly always suspected from the vascular pattern occupying the environs of the serpentine channel. Persons not familiar with this entity may have falsely interpreted this as a giant saccular aneurysm or arteriovenous malformation surrounded by an intracerebral hematoma. It was, however, the globular thrombosed portion of the aneurysm that caused the "mass effect."

Discussion

These aneurysms have most often shown evidence of an expanding mass effect rather than of a subarachnoid hemorrhage. They occur as globoid masses involving the middle cerebral artery and cause their neurological picture by at least two mechanisms: local pressure and distal ischemia. A considerable volume of blood flows through these aneurysms and usually supplies a region of temporoparietal cortex. The lesions may enlarge to some extent, but it is the progressive thrombosis within the lumen which largely accounts for their mass effect. With progressive narrowing of the vascular channel, regions of brain previously well supplied may become ischemic. The rapidity of the narrowing determines whether ischemia or infarction occur. Reports in the literature have documented the serious deficits resulting from total thrombosis of the aneurysm.7,8 Thrombosis may occur spontaneously or may be the result of inadvertent operative violation of the internal channel. In some cases, the development of transcortical collaterals may compensate for slowly progressive narrowing of the lumen.8

Evidence for edema being a deleterious result of the presence of the aneurysms is gathered from clinical observation in our second patient who improved significantly pre-operatively when placed on a course of steroids. His aphasia and hemiparesis improved preoperatively within 12 hours of the institution of dexamethasone therapy.

Jane1 reported a case similar to those presented here in which a 1-year-old child was discovered at postmortem examination to be harboring a giant aneurysm fed by the posterior inferior cerebellar artery. The child did not die as a direct result of the undisturbed aneurysm, and it therefore seems evident that giant aneurysms can be of congenital origin and be asymptomatic during childhood.

In the past, surgical excision of these lesions has been associated with considerable morbidity (Table 1); however, preoperative recognition of its anatomic and pathophysiological nature greatly facilitates the operative plan. Clipping the feeding vessel and avoiding inadvertent entrance into the vascular lumen are major points to be recommended.

There have been speculations in the literature as to the etiology of these lesions. Some authors prefer to regard these giant masses as expansions of smaller classical saccular aneurysms.1,2,5,6 Since the majority of these lesions occur in adulthood there seems adequate time for expansion to occur; however, it is unusual that the enlargement that has taken place has not been accompanied by signs and symptoms of subarachnoid hemorrhage. From a study of the literature and evaluation of the existing clinical material on the subject, it seems evident that these lesions represent a significantly different pathological process from that of the typical saccular aneurysm. Both the clinical and pathological differences between our cases and those of classical saccular aneurysms seem to warrant their differentiation into separate categories.

The neuroradiological picture is so distinct it seems fitting that the aneurysms should be re-named in the literature on the basis of their angiographic picture. We therefore propose that these entities be recognized as "giant serpentine aneurysms."

References


J. Neurosurg. / Volume 46 / January, 1977

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