Approximately 20% of all clinically significant intracranial aneurysms arise from the MCA. The majority of these are saccular, and typically originate as a direct extension of the main MCA trunk (M₁ segment) between and beyond its two major branches near the genu. Some MCA aneurysms, however, exhibit a fusiform (spindle-shaped) appearance, at least when they are small and when viewed externally. Fusiform aneurysms have different underlying pathological features, hemodynamics, anatomical distributions, natural histories, and treatments than do the saccular variety.

We have reviewed all reported cases of fusiform lesions that arose from the MCA, and we eliminated those associated with an obvious underlying cause (that is, infection, trauma, and so on). The remainder, herein referred to as spontaneous fusiform MCA aneurysms, were presumed to be caused by dissection, atherosclerosis, or some unknown factor. This review included only those cases in which the clinical course and arteriographic features could be clearly extracted; the remainder were discarded from analysis because of insufficient data. To the 62 cases identified in this way we added 40 of our own that were encountered in our clinical practices during the last two decades.

An analysis of this group of patients provides an understanding of the cause, clinical spectrum, natural history, and treatment of these lesions that we believe is applicable to similar intracranial fusiform aneurysms at other locations.

Anatomy and Terminology

Anatomy of the MCA

The MCA begins at the terminal bifurcation of the ICA and supplies the lateral two thirds of the cerebral hemisphere centered around the sylvian fissure (Fig. 1). The vessel initially runs laterally, paralleling the sphenoid ridge within the sphenoidal compartment of the deep sylvian fissure. At the level of the limen insula, the MCA makes an abrupt turn (genu) posteriorly and superiorly to enter the...
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deep insular compartment of the sylvian fissure. Branches from these insular trunks then extend across the operculum of the sylvian fissure to reach the cerebral cortex.

The MCA can be divided into four segments as follows: 1) M1, the segment between the CA bifurcation and the genu; 2) M2, the segments that run over the deep insular surface; 3) M3, the segments that span the opercular surface of the sylvian fissure; and 4) M4, the cortical branches. The M segment typically divides into superior and inferior trunks just proximal to the genu; these vessels then run in the deep insular compartment of the sylvian fissure. The M segment gives rise to lenticulostriate vessels from its posterosuperior surface, and often contributes an early or anterior temporal branch off its anteroinferior surface. Recurrent lateral lenticulostriate branches occasionally originate from proximal portions of the superior and inferior trunks.76

Inclusion Criteria

Intracranial aneurysms can be classified according to their shape and origin into saccular (sometimes called berry) and nonsaccular types. Saccular aneurysms typically arise at bifurcations, along a curve of the parent vessel, and point in the direction in which the flow would have proceeded if the curve had not been present.54,55 These lesions typically arise between two exiting branches as a consequence of hemodynamic stress or increased flow at a bifurcation site and/or accentuated turn in the vessel. The resultant lesion resembles a blind pouch, sac, or berry attached to the bifurcation site by a stalk (or neck). These aneurysms receive the full force of the pulsatile flow from the parent artery directly into their fundus, and patients often present with SAH.

Nonsaccular aneurysms do not typically originate from bifurcations, but arise at a point at which the vessel wall breaks down from external trauma or weakening caused by infection, inflammation, neoplasia, atherosclerosis, or spontaneous dissection.20 The resultant defect, when subjected to infection, inflammation, neoplasia, atherosclerosis, or spontaneous dissection. These lesions typically arise between two exiting branches as a consequence of hemodynamic stress or increased flow at a bifurcation site and/or accentuated turn in the vessel. The resultant lesion resembles a blind pouch, sac, or berry attached to the bifurcation site by a stalk (or neck). These aneurysms receive the full force of the pulsatile flow from the parent artery directly into their fundus, and patients often present with SAH.

All nonsaccular fusiform MCA aneurysms associated with trauma, infection (myotic), inflammation, and neoplasia have been excluded from our review, because each has an obvious origin contributing to its formation and subsequent clinical course. The remaining lesions, including those thought to be due to spontaneous dissection, atherosclerosis, or unknown causes, are herein referred to as spontaneous fusiform MCA aneurysms and comprise the clinical material for this report.

Specific Lesion Characteristics

Spontaneous fusiform MCA aneurysms are classified in this study according to the following criteria: 1) site of origin (location), including proximal (M1 segment proximal to the genu), insular (M2 segments deep within the insular compartment of the sylvian fissure, beginning at or beyond the genu), and distal (M3, or M4 segments) categories; 2) size, defined by maximum external diameter or length of vessel segment involved (small, < 10 mm; large, 10–24 mm; and giant, ≥ 25 mm), as viewed at surgery, postmortem examination, on CT and/or MR imaging studies, or, when other methods of measurement were not available, by arteriography; and 3) luminal shape.

All aneurysms exhibited a spindle shape when viewed externally, with entering and exiting branches present on somewhat opposite sides of the fusiform enlargement. The arterial lumina of these lesions varied substantially, however, and a particular subtype was classified as serpentine, which was used to denote cases in which the diseased lumen extends longitudinally along the original artery’s major axis and curves, creating a seemingly wandering or serpentine course. Other classifications were as follows: focal dilation, in cases with a dilated enlargement of the vessel lumen without focal stenosis; and stenosis or occlusion, when a focal reduction or loss of the vessel lumen was present.

Fig. 1. Drawing showing MCA anatomy terminology, and the distribution of case material. Aneurysms arising from M1 are termed “proximal,” those having their origin on M2 are “insular,” and those originating from M2 or M3, are “distal.” The M1 is the segment between the CA bifurcation and the genu; M2 are the segments that run in the deep insular compartment of the sylvian fissure, including the superior and inferior trunks; M3 are the segments and branches that traverse the opercular surface of the sylvian fissure to reach the cortical surface; and M4 are the cortical surface branches. A. = arteries; ETA = early or anterior temporal artery; N = number; Tr = trunk.

Presentations, Treatment, and Outcomes

Clinical Presentation. The clinical presentation was based on symptoms at the time of diagnosis. Categories include the following: 1) incidental or asymptomatic, discovered during workup for unrelated symptoms; 2) nonspecific, headache without hemorrhage or other neurological signs or symptoms; 3) ischemia, TIAs, or completed stroke; 4) mass effect (with or without seizures); and 5) hemorrhage, subarachnoid and/or intraparenchymal. The clinical presentations were assigned somewhat arbitrarily, because many patients experienced both ischemia and mass effect, or presented with seizures and also had some ischemic changes. Hemorrhage was also sometimes difficult to determine; it was often obscured by the suddenness of an ischemic event or by a large lesion size with calcification and hemosiderin in its margins. In such cases, we chose the most obvious
presenting feature as judged by the clinical information available.

Treatment Categories. Treatments were divided into categories principally based on intraaneurysmal blood flow reversal, and included the following: 1) none—observation only; 2) minimal—exploration, “debulking,” wrapping, or clip occlusion without parent vessel sacrifice; 3) intermediate—flow reversal alone by using proximal occlusion, or trapping with or without resection; and 4) maximal—flow reversal, proximal occlusion, or trapping (as long as the lumen of the entering vessel was still patent) with or without resection, combined with some form of an internal or external bypass procedure designed to augment distal MCA blood flow.

Outcome Scale. An outcome scale was devised to analyze disease and treatment responses. Patients were assigned a good grade if they could return to most or all of their pre-disease activities after treatment, either by improvement or resolution of presenting symptoms, including hemorrhage or stroke, or by stabilization of a preexisting deficit at a functional level. Poor outcomes were assigned to patients with major neurological deficits or death. Many patients exhibited significant neurological deficits before treatment was instituted, which accounted for many of the poor outcomes. Outcomes were also classified according to whether the treatment was associated with improvement, stabilization, or worsening of presenting signs and symptoms, either immediately or over time, after the procedure was used.

Clinical Material and Methods

Patient Population

Forty cases encountered in our departments during the last two decades were combined with 62 similar cases identified from the English language literature published between 1950 and 2000 in which there was sufficient data to allow comprehensive analysis.\(^1\) Among these, 97 presented with dissecting-type lesions; 3 were excluded because data were not available; it does not include five cases with atherosclerotic lesions. The male/female ratio was 1.3:1 (53 male, 41 female patients); the mean age at presentation was 36 years (range 19 days–76 years).

Age and Sex Distribution

The age and sex distribution of the dissecting type of aneurysm (97 patients) is summarized in Fig. 2. The data represent the patient’s age at the time of symptom onset or initial diagnosis, and ranges from 19 days to 76 years (mean 36 years). The majority of patients (59%) were younger than 40 years of age; 53 were male and 41 were female (1.3:1 male/female ratio). Three were excluded because data were unavailable.

Results

Neuroimaging Features

Figure 3 depicts examples of the characteristics identified on neuroimages.

Small Lesions. Arteriography might demonstrate either focal MCA stenosis/occlusion or dilation. The lumen of such lesions can be difficult to distinguish from atherosclerotic or vasospasm-related narrowing. Magnetic resonance imaging and MR angiography were essential in demonstrating focal external dilation of the affected MCA segment, with perhaps some adjacent hemosiderin staining reflecting old hemorrhages.

Large Lesions. In most large lesions there was dilation of the affected segment and some, especially the recently symptomatic ones, were accompanied by perilesional edema. The CT scans might demonstrate acute hemorrhage or intraluminal thrombosis, and with contrast enhancement, the aneurysm itself and any adjacent ischemia could be seen. Arteriography clarified the segmental MCA dilation, which usually at this stage had good flow distally into peripheral MCA branches. Magnetic resonance imaging and MR angiography effectively demonstrated the fusiform external shape, focal luminal dilation, any associated ischemic changes in peripheral MCA branches, and mural thrombus causing secondary changes in the vessel lumen.

Giant Lesions. Most lesions had encountered a major bend of the MCA by the time they were this size, and the combination of vessel curvature and partial intraluminal thrombus imparted a serpentine shape to the MCA lumen. Both CT scans and MR images demonstrated well the lesion and adjacent parenchymal edema from ischemia or fo-
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Fig. 3. Neuroimaging studies demonstrating aneurysm size as judged by maximum external diameter, lumen variations as revealed by angiography, and other characteristics. A–C: Small lesions (< 10 mm); the luminal appearance at this size includes stenosis or occlusion (focal reduction or loss of vessel lumen diameter), or focal dilation. Axial T2-weighted MR image (A) demonstrating focal dilation of the proximal MCA, with adjacent hemosiderin staining reflecting old hemorrhage. Magnetic resonance arteriogram (B) obtained in the same patient, confirming focal MCA dilation of the distal M1 segment. An anteroposterior (AP) arteriogram (C) obtained in another patient, revealing M1 stenosis, with the bulge on the inferior vessel wall reflecting focal weakness. This abnormality would be difficult to distinguish from atherosclerotic luminal narrowing, but an MR image revealed that the external vessel diameter widened into fusiform shape. D and E: Large lesions (10–24 mm); at this size, most lesions exhibited focal dilation of the vessel lumen without focal stenosis. Mural thrombus often causes secondary changes in the vessel lumen or peripheral branch ischemia. Contrast-enhanced CT scan (D); note adjacent ischemic edema and mass effect. An AP arteriogram (E) obtained in the same patient, demonstrating good flow distally into peripheral MCA branches. F and G: Giant lesions (≥ 25 mm); most will have encountered a major bend of the MCA by the time they are this size. As the dissection extends longitudinally along the MCA, the combination of the vessel curvature and the partial intraluminal thrombus imparts a serpentine shape to the MCA lumen. The often recanalized lumen may no longer be lined with vascular endothelium. Axial MR image (F) demonstrating a giant aneurysm with adjacent parenchymal edema from ischemia and/or focal compression. Note the surrounding hyperintensity indicative of hemosiderin deposition; also note the irregular lumen “wandering” through the anterior portion of a large intraluminal thrombus. An AP arteriogram (G) demonstrating a typical serpentine channel. Note the MCA terminal branches exiting the diseased MCA segment, with filling delayed by the combination of the circuitous channel and accompanying partial luminal obstruction. H and I: Atherosclerotic variant. This type typically occurs in older individuals, and has a disproportionate degree of atherosclerosis and calcification in the vessel wall. Contrast-enhanced CT scan (H) demonstrating elongated proximal MCA dilation in 60-year-old man presenting with SAH. Arteriogram (I) obtained in the same patient. Surgical exploration identified a focal “bleb” off the surface of a very atherosclerotic vessel that appeared to be an extension of a dissection. The site of focal weakness was treated with clip occlusion, and the remainder of the diseased artery was left intact. Note that when viewed externally, all lesions exhibit fusiform characteristics.
Aneurysm Location, Size, and Luminal Shape

Aneurysm location, size, and luminal shape distributions are outlined in Fig. 4. Overall, 67 (69%) originated proximal to the MCA genu, 21% were insular (Mg, segment), and 10% were distal (Mg or M, branches). Twelve lesions were small (12%), 19 were large (20%), and 66 were giant (68%). Luminal stenosis or occlusion of the MCA was seen in 10 patients (10%), focal luminal enlargement was identified in 54 (56%), and a serpentine channel was found in 33 (34%); all but one of the serpentine lesions were giant. The arteriographically identified lumen size or shape was an inaccurate predictor of an individual lesion’s true size, shape, or associated mass effect.

Presentation and Evolution of Symptoms and Neuroimaging Features

The clinical history was unavailable, ambiguous, or did not allow definitive categorization of clinical presentation in two patients, leaving a total of 95 cases for analysis. Of these, 21 had incidental and asymptomatic lesions, 86% of which exhibited focal dilations of the MCA lumen. None of the stenoses/occlusions was asymptomatic, and giant serpentine lesions were rarely so.

When they were symptomatic, small, large, and giant aneurysms with focal dilations of their lumen had SAH rates of 80, 62, and 23%, respectively, whereas serpentine aneurysms had a hemorrhage rate of 14% (Fig. 5). Thirty-one percent of focal dilations and 69% of serpentine aneurysms presented with symptoms related to mass effect. Ischemic symptoms (TIA or completed stroke) were the presenting feature in 31% of aneurysms with focal dilations, and in 17% of serpentine aneurysms. Recognizing the somewhat arbitrary separation of the latter two categories (ischemic symptoms and mass effect), the combined presentation rates were 20, 38, and 77% for small, large, and giant focal dilations, respectively, and 88% for serpentine lesions.

Changes in the aneurysm size, shape, or luminal characteristics occurred in 16 patients before definitive treatment was implemented, with intervals between presentation and treatment ranging from 3 weeks to 5 years (mean 18 months), providing an opportunity to assess the natural history of these lesions (Fig. 6). In one patient angiographic findings normalized 1.3 years after presentation with headache and a 2.5-cm focal dilation. Another lesion presenting as SAH changed from a focal stenosis to a dilation over a period of 2 months. In five others an established focal dilation enlarged during a period of several months; four aneurysms presenting as focal dilations evolved into giant serpentine lesions within time intervals spanning 2 to 5 years. Three patients experienced spontaneous thrombosis of giant lesions several months after diagnosis, including two in whom this was associated with acute MCA stroke.

Pathological and Surgical Findings

When described in detail, surgical exploration, particularly in larger and giant lesions, invariably demonstrated hemosiderin staining in the adjacent cortex, indicating remote prior hemorrhage. Significant encephalomalacia was seen in many cases, consistent with longstanding mass effect and ischemic damage. Atherosclerosis was an inconsistent finding, but calcifications within the walls were common in longstanding lesions. In giant serpentine lesions, a currant-jelly consistency was often described in the periluminal thrombus, and the residual lumen was seemingly poorly endothelialized and obviously friable in nature.
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Treatment and Outcomes

The results of treatment and the ultimate clinical outcomes are summarized in Fig. 7. Patients receiving treatments that led to intraaneurysm blood flow reversal had better outcomes compared with the untreated and/or nonreversed groups. Those treated with flow reversal and a patent accompanying distal bypass had the best results, considering that such treatments were usually performed in patients harboring the largest and most complicated lesions.

Generally, in lesions described in the literature as case reports, especially those treated in the “bypass era,” there were good outcomes, and these reports were published to validate the use of aggressive treatment. One can presume that a single treated case with a poor outcome would not initiate a published report or description unless there was something unique about its presentation or pathological features.

Reasons For Poor Outcomes

Only three reports (including this one) describe a series of cases in which a more comprehensive view of the incidence and cause of surgical complications and poor outcomes can be at least inferred.6,10 In general, poor results came about for one of the following reasons: 1) delay in intervention until a symptomatic patient experienced further aneurysm growth and ultimately an irreversible insult; 2) failure of a functional bypass; or 3) side-wall clip occlusion of the lesion with subsequent regrowth.

Delay in Intervention. The case illustrated in Fig. 6C–E represents an example of delay in intervention with a subsequent poor outcome, despite eventual aggressive intervention. This 71-year-old woman presented with transient left hemisphere dysfunction that resolved over several weeks. Her initial neuroimaging studies demonstrated a giant focal dilation of the M1 segment. Because of her age and essentially normal neurological status, a conservative approach of continued observation was chosen. Eleven months later, she presented again with a rapidly progressive, marked left hemisphere deficit and a near doubling of the aneurysm size, now with restricted distal MCA flow. Despite emergent trapping, resection, and bypass, she did not recover. Intraoperative pathological studies showed fresh thrombosis within the aneurysm obstructing MCA outflow and obliterating several nearby exiting branches.

Failure of Bypass. Another patient with a distal MCA lesion underwent resection and reanastomosis of the entering and exiting branches. She did well clinically, despite the fact that a postoperative arteriogram revealed that the anastomosis between branches of the MCA was not patent. Six months later, she experienced a significant hemorrhage in the posterior sylvian region from tiny leptomeningeal vessels that had dilated in response to the focal ischemia produced by the resected vessel segment. Presumably, a well-functioning connection would have eliminated the need for such channels.

Regrowth After Clip Occlusion. Three aneurysms exhibited regrowth after surgery in which the “aneurysmal” portion of the lesion was treated with clip application while sparing the orthograde MCA luminal flow. In these postoperative cases, the focal dilation recurred over a period of days or weeks, indicating that residual vessel wall weakness persisted.

Asymptomatic Aneurysms

Of the asymptomatic aneurysms, a category that includes 21 lesions discovered incidentally or during an evaluation of headaches, 86% were focal dilations, 14% were serpentine, and none were due to occlusions or stenosis. The location of these lesions was proximal in 57% of cases, insular in 38%, and distal in just 5%; the size was large in 43%, giant in 38%, and small in 19%. Two patients with asymptomatic fusiform aneurysms suffered an SAH from another, saccular lesion. Two of the asymptomatic cases were observed to evolve during follow up. One patient in whom enlargement of a focal dilation occurred during a period of 6 weeks was treated with resection and bypass, and had a good outcome. The other patient, who initially underwent only exploration of a giant, serpentine lesion, did well for...
Fig. 6. Evolution of lesions over time. Changes in aneurysm morphological features or size during the period of follow up (before or after treatment) were viewed as snapshots in the stages of evolution of these lesions. One patient progressed from stenosis to focal dilation, five exhibited enlargement of their focal dilation, three lesions evolved from focal dilations into serpentine aneurysms; one focal dilation regressed to a normal angiographic appearance.
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13 months, but then presented with MCA thrombosis at the proximal aneurysm origin and died of cerebral infarction. No treatment was given in nine of the 21 asymptomatic cases, and eight of these patients had a good outcome after a median follow-up period of 13 months. One patient died of an SAH involving another saccular aneurysm.

Of those treated, five (42%) underwent minimal therapy (wrapping or direct surgical clip occlusion without parent vessel sacrifice), five (42%) underwent maximal therapy (proximal occlusion or trapping, with or without resection, combined with a bypass procedure), and two (16%) received intermediate therapy (proximal occlusion or trapping with flow reversal, without bypass). Four (80%) of the five treated with maximal therapy had good outcomes, whereas two (40%) of the five treated with minimal therapy had good outcomes.

Discussion

We propose that spontaneous fusiform MCA aneurysms usually develop as a result of arterial dissection. The process of dissection may be multifactorial and may include both congenital and acquired factors. Most dissecting aneurysms involving the cranial vasculature arise from the extracranial cervical CA or VAs near the C1–2 level. Intraluminal dissections most commonly involve the posterior circulation and, especially affect the VA3,33,69 and basilar artery11,28,77,78 in older patients. Dissecting aneurysms involving the

and three giant aneurysms thrombosed spontaneously. Three focal dilations treated with aneurysm clip application without parent vessel sacrifice exhibited regrowth after surgical obliteration. A and B: Progression from stenosis to focal dilation in a 41-year-old woman presenting with SAH. Arteriogram (A) demonstrating focal narrowing of a peripheral MCA branch (arrow). Repeated arteriographic study (B) revealing rehemorrhage several days later that required emergency hematoma evacuation. Repeated arteriography performed 2 months later demonstrated an 8-mm focal dilation of the artery at the same site as the prior stenosis. Reprinted from J Neurosurg 74:504–507, 1991. C–E: Neuroimaging revealing enlargement of focal dilation in a 71-year-old woman presenting with a single reversible episode of expressive aphasia and right hemiparesis. The initial MR image (C) and arteriogram (not shown) demonstrated giant focal dilation (40 mm) of the M1 segment; the patient remained asymptomatic without intervention. Repeated MR image (D) obtained 11 months later because of rapidly progressive fixed deficits, demonstrating growth of the lesion to 70 mm. Arteriogram (E) demonstrating giant focal dilation with partial outflow obstruction to peripheral MCA branches. Other cases demonstrated similar progressive growth, generally within 1 to 2 years. F–I: Arteriograms revealing progression from focal dilation to serpentine channel in a 37-year-old man whose neuroimaging studies demonstrated growth and evolution of 15-mm focal dilation into a 40-mm serpentine lesion over a 2-year period. Two other patients had similar angiographically confirmed evolutions. Oblique AP (F) and lateral (G) arteriographic views obtained at presentation. Same views (H and I) obtained 2 years later. J–L: Regrowth of lesion after surgical clip application in a 23-year-old man presenting with SAH. Initial arteriographic study (J) demonstrating a fusiform lesion of the superior trunk. Early postoperative arteriogram (K) demonstrating excellent aneurysm obliteration. Second arteriogram (L) obtained after recurrence of SAH 3 weeks postsurgery. Note regrowth of aneurysm from the remaining patent vessel, which had been diffusely weakened by the initial dissection.

intracranial anterior circulation most commonly arise from the MCA, with a significant incidence in children and adolescents.11,31,59,82

Our study confirms previous reports that spontaneous fusiform aneurysms in the anterior circulation, especially those involving the MCA, are more often found in younger patients.25,32,36,43,62 The age of patients presenting with these lesions (mean 36 years) contrasts sharply with that of patients with aneurysms in the posterior circulation, those with saccular MCA aneurysms, and those with our atherosclerotic variant, all of which peak in incidence at least a decade later.43,52,80 Male predominance, which was well demonstrated in the series reported by Drake and Peerless,18 is also corroborated in our material. These factors, along with the finding that some lesions demonstrate dynamic, serpentine evolution on neuroimaging studies, as in Fig. 6, support arterial dissection as the origin of these aneurysms.

Intracranial arterial dissection has been associated with other congenital vascular processes such as ICA redundancy, moyamoya disease,79 vertebrobasilar junction fenestration,83 fibromuscular dysplasia,49 and α1-antitrypsin deficiency.63 Some arterial dissections have also been reported within the same family, suggesting a hereditary influence.61 Most lesions, however, exhibit no obvious identifiable pathological cofactor, other than atherosclerosis in older patients.

Pathological studies have confirmed that intracranial arterial dissections are frequently (if not invariably) associated with intramural hemorrhage. In MCA dissections that have been pathologically described, the intramural hemorrhage was observed between the elastica and media (that is, a subintimal dissection), which often produced luminal stenosis or occlusion.66,51,65,70 In contrast, approximately half of pathologically investigated intracranial VA dissections were associated with intramural hemorrhage between the media and adventitia,20 with consequently higher potential for clinically significant rupture into the subarachnoid space.

Four of our cases arose in the patient’s 1st year of life. Pathological examinations in two of these cases showed an aneurysm wall composed of a thin layer of fibrous tissue without an elastica interna. Considering the age at presentation and lack of prior injuries in these cases, it is highly un-
likely that acquired or environmental factors played a significant role in aneurysm formation. In older individuals, however, there is evidence to suggest that atherosclerosis is a predisposing or associated cofactor. 1,9,16,25

Fusiform aneurysms of the intracranial VA and ICA frequently present with hemorrhage, and in this respect are substantially different from such lesions in the MCA. The available data do not explain this difference. One possible explanation is that, in arteries that are tethered by their dural attachment just proximal to the dissection site (such as the ICA or VA), the pulsatile and expansive forces can only be dissipated by vessel wall failure, leading to a higher incidence of clinically significant hemorrhage. In arteries in which there is no tethering of the perilesional vessel, the pulsatile and expansive forces in a growing dissection may be dissipated by elongation and the development of ectasia, with gradual enlargement of the dissecting segment resulting in the formation of a giant or serpentine aneurysm.

The large number of proximal dissections in younger people indicates that the M1 segment may have some underlying embryological defect or other structural abnormality that further predisposes that region to arterial dissection. A similar explanation could also apply to VA lesions that arise just distal to a transition zone where the vessel is changing from an extracranial to intracranial artery, usually proximal to the origin of the posterior inferior cerebellar artery.

Patients with lesions in the atherosclerotic category were older and had evidence of atherosclerosis in other organ systems. The dissections in this group are presumably related to the hazards of atheroma exposed to high-pressure arterial blood flow and its attendant increased shear stresses. Such secondary changes are not specific to the MCA, but can occur in any intracranial vessel affected by atherosclerosis.

Natural History

We have developed a proposed sequence of aneurysm evolution that correlates with the pathological findings and the visible progression of the lesions outlined in Fig. 6. As proposed in Fig. 8, the initial pathological event in the formation of a spontaneous MCA fusiform aneurysm is a dissection accompanied by intramural hemorrhage within the vessel wall. Outward extension of the injury may facilitate SAH, whereas confinement of the intramural thrombus between the dissected layers may promote luminal narrowing or occlusion. Rupture into the arterial lumen may release embolic material into the distal MCA. If flow is reestablished, the weakened vessel wall may stabilize or focally dilate. Progressive focal dilation of the affected segment, when combined with longitudinal extension of the dissection and varying degrees of intraluminal thrombosis, eventually evolves into a serpentine aneurysm. Intraluminal thrombosis may embolize distal channels. The origins of branches arising along the dissected segment may become occluded by the thrombus or by longitudinal advancement of the aneurysm as it grows.

The clinical features closely correlate with the morphological features of the given aneurysm. The exact likelihood and timing of pathological evolution for an individual lesion, however, is still not certain. Some hemorrhage probably occurs during aneurysm formation, but can generally be characterized as a leakage rather than a frank rupture for most lesions. Most patients with dissections will exhibit some hemosiderin staining in the adjacent subarachnoid space and gyri. In intradural VA dissections, the weakened arterial wall remains prone to hemorrhage for at least several weeks after formation, after which the bleeding risks appear to lessen progressively. 20 A similar natural history was not identified in the MCA lesions described here.

In contrast to VA lesions, the initial MCA dissection is usually clinically silent unless it is associated with ischemia from luminal obstruction. Varying degrees of intramural or intraluminal thrombosis or an intimal flap may produce symptomatic stenosis or occlusion with accompanying TIA and/or stroke, particularly in small, newly formed lesions. Such sequelae may be one explanation for hemiplegia of childhood, and for some older patients presenting with ischemic symptoms from atherosclerotic MCA stenosis.

Some (perhaps many) MCA dissections presumably resolve spontaneously, according to both clinical and neuroimaging evidence. Assuming that blood flow is reestablished in the affected arterial segment, however, the damaged and weakened wall may promote focal dilation of the lumen beginning at the point of arterial injury. Once formed, the MCA lumen may remain stable at that size, or it may expand both laterally and longitudinally to create larger focal dilations or serpentine lesions. As the dissection enlarges, the risk of hemorrhage progressively diminishes, especially when the lesion has assumed a serpentine shape with substantial intraluminal thrombosis. Simultaneously, however, the risks increase for ischemia from embolization, parent vessel thrombosis, or occlusion of the orifices of branches exiting from the affected segment. Once symptoms of mass effect or ischemia appear, the natural history appears to be progressive and poor. Lesion expansion usually continues, and the distal MCA territory is under constant threat of further ischemic damage.

Treatment Protocols

Management strategies should be based on the presence and type of symptoms, the lesion size and location, and the risks accompanying intervention. In our opinion, most small and some larger focal dilations, especially those that are asymptomatic, should be treated conservatively unless serial neuroimaging assessments indicate significant enlargement over time. The outcomes in the asymptomatic group whose lesions were managed nonsurgically support this strategy, although the duration of follow up and the number of patients were limited. Once symptoms appear, however, aggressive intervention is warranted.

Stenotic or occlusive lesions, particularly from the atherosclerotic variant, which present with acute ischemic symptoms should be treated with management strategies aimed at limiting the extent of stroke. The use of such modalities as anticoagulation therapy, intraarterial thrombolysis, and endovascular intervention must obviously be weighed against the risks of precipitating a hemorrhage. Lesions presenting with bleeding are fortunately uncommon, and should be treated with surgical or endovascular obliteration of the bleeding source combined with maintenance of distal MCA flow.

Symptomatic large or giant lesions presenting with subacute or chronic ischemia should be treated with flow reversal (direct clip application or endovascular luminal oblit-
Fig. 8. Drawings showing the origins and stages of evolution of MCA dissections. Arrows indicate direction of blood flow. A: Arterial dissection, due to congenital or acquired causes, with intramural hemorrhage between the intima and media, producing demonstrable focal narrowing of the peripheral MCA branch. B: Extension of bleeding (arrows) into subarachnoid space, common but rarely clinically significant, especially when arising from the M₁ segment. C: Rupture into the true lumen, with potential for distal embolization. D: Further expansion of intramural clot leading to vessel occlusion. E: Recanalization or expansion of lumen, creating focal fusiform dilation of affected MCA segment. F: Progressive enlargement of dissection both laterally and longitudinally. G: Serpentine channel forms as disease extends longitudinally and incorporates vessel curves, combined with varying degrees of intraluminal thrombosis.
etration of the MCA just in front of the aneurysm origin) combined with a bypass procedure to protect the distal circulation. The gradually restricted flow that occurs as a serpentine lumen evolves often allows the leptomeningeal collateral vessels time to develop, making acute parent vessel sacrifice safer than for less mature lesions in which the channels have not had the need or the time to dilate effectively. Collateral circulation beyond the point of vessel sacrifice, however, is dependent on leptomeningeal channels that are unpredictable in their ability to sustain and protect the cortex. The vessels at greatest risk of intraoperative injury are the lenticulostriate arteries, because they are end arteries that have no significant intercommunication with other vessels. In our opinion, their survival and that of the cortical branches should not be left to chance, and, whenever possible, a patent bypass offers the best method for their protection.

The choice of an arterial as opposed to a venous bypass conduit is dependent on the size of the donor arteries available and the site and size of the MCA territory affected by the diseased vessel to be sacrificed. In this series, the best outcomes occurred in the maximally treated group, in which a combination of flow reversal and a patent bypass conduit was used. For more peripheral lesions, the superficial temporal artery is the ideal donor source, whereas a large vein or radial artery is generally recommended when the anterior temporal artery is the ideal donor source, whereas a large vein or radial artery is generally recommended whenever a lesion is encountered in the M1 segment. Anson, et al., in a series of 13 mostly giant fusiform MCA aneurysms, reported generally good results with this strategy. Drake and Peerless, in a similar series of 12 patients, described good or excellent outcomes in 88%, and, just as importantly, poor results in 75% of patients who did not receive functioning bypasses. Intraoperative angiography is helpful in determining graft patency, and in all patients undergoing bypass procedures, antiplatelet therapy is initiated immediately postoperatively to reduce the risks of graft thrombosis.

Distal blood flow can be successfully managed in some patients by performing resection of the affected arterial channel and an internal bypass procedure, in which either a primary reanastomosis or an end-to-side union with an adjacent patent MCA trunk is used. When dealing with direct approaches that preserve blood flow in the affected MCA branch, however, it must be remembered that the injury associated with dissection is invariably a circumferential one. Direct "aneurysm only" clip application is associated with a high rate of regrowth of the lesion, particularly if a right-angled fenestrated or parallel clip preserving flow in the normal half of the affected vessel is applied (Fig. 6J–L). This phenomenon was observed in three cases in our series, despite seemingly excellent intraoperatively confirmed aneurysm obliteration. Reexpansion of the weakened residual arterial channel occurred in two cases within a matter of days, and all patients ultimately required additional surgery.

Trapping is invariably necessary for lesions presenting with mass effect; this is performed to control bleeding once aneurysm debulking and resection have been completed. After thorough internal debulking and inspection for patency of exiting channels, the aneurysm wall should be left largely in place so as not to affect leptomeningeal channels that may be adherent. Small exiting trunks can be anastomosed to each other and then connected to a single bypass source that effectively irrigates all distal circuits. The proximal MCA must be reconstructed with clips in such a way as to ensure that the origins of the lenticulostriate vessels are preserved and receiving orthograde flow from the ICA or retrograde flow from the bypass.

The development of intracranial stents may prove very useful in managing these lesions. Similar devices already have a clear role in the management of extracranial dissections. When lenticulostriate artery origins are free of the aneurysm, endovascular proximal MCA obliteration may be used to avoid a deep dissection within the sylvian fissure to find the proximal blood supply to the lesion. This technology is not, however, a substitute for the collateral circulation protection provided by a judiciously and skillfully applied bypass procedure.

Conclusions

Based on the spectrum of clinical, pathological, neuroimaging, and intraoperative findings, two distinct categories of spontaneous fusiform MCA aneurysms appear to represent variations of arterial dissection. As reported here, the evolution of these lesions over time likely models that for most fusiform intracranial aneurysms encountered elsewhere. Most MCA lesions arise from the M1 segment and start as a spontaneous intramural hemorrhage from a seemingly healthy portion of the vessel unassociated with significant atherosclerosis. Once formed, the earliest visible stage is a focal stenosis or occlusion of the affected portion of the vessel, which can be associated clinically with SAH or ischemic symptoms. If the lumen remains patent, the diffusely weakened arterial injury may dilate, and the dissection may then enlarge further. Bleeding risks eventually drop to low levels, but the formation of intraluminal thrombus and the longitudinal extension and growth of the lesion places the distal MCA territory at increasing risk for mass effect and ischemia as the lesion enlarges. Aggressive intervention is indicated for symptomatic lesions, with a primary aim of achieving flow reversal in the aneurysm while preserving blood flow to the distal MCA territory.

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Dedication

Dedicated to the memory of our friend Chris Gaposchkin, M.D., Ph.D., who worked diligently on this manuscript, and who was an inspiration to us all.

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Manuscript received May 1, 2002. Accepted in final form March 21, 2003. Address reprint requests to: Arthur L. Day, M.D., Department of Neurosurgery, Harvard University School of Medicine, Brigham and Women’s Hospital, 75 Francis Street, Boston, Massachusetts 02115. Email: aday1@partners.org.