Chapter 5
Brain Vascular Disease

Brain Aneurysm: Ruptured

Brain aneurysms, which may be single or multiple, usually occur as an isolated phenomenon, but rarely can be associated with polycystic kidney disease (a genetic disorder), brain AVMs, or endocarditis.

A brain aneurysm that has ruptured is at high risk for re-rupturing. The re-rupture rate is about 20% in the first 2 weeks and about 50% in the first 6 months. Furthermore, some of the treatments for vasospasm, such as hypertensive therapy and intra-arterial papaverine, are much riskier if the aneurysm is not secured. For these reasons, a ruptured aneurysm in an otherwise viable patient should be treated in a timely fashion, ideally within 24 h of presentation.

Brain aneurysms can be treated by endovascular "coiling" or by craniotomy and "clipping." The coiling procedure is much less invasive and morbid and therefore should be the procedure of choice in almost all cases. The randomized controlled ISAT trial of 2143 patients showed that patients with ruptured aneurysms had significantly better clinical outcomes when they underwent endovascular coiling of aneurysms instead of craniotomy and clipping [21]. In the rare case in which a ruptured aneurysm cannot be coiled, clipping would be appropriate. When an anterior circulation aneurysm must be clipped, the use of neuronavigation can help avoid the frontal sinus, which is well worth avoiding if possible (in this and other frontal craniotomies). Technical keys to these cases, when needed, are early establishment of proximal arterial control and, upon clipping, preservation of parent vessels.

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Brain Aneurysm: Unruptured

The indications for intervention are much less clear here. For most unruptured aneurysms, the annual risk of hemorrhage is likely around 0.05–0.1% per year (ISUIA study, 2621 patients at 53 centers [22]). This is consistent with the known rate of brain aneurysm in the population of about 3–5% and the known annual rate of brain aneurysm rupture. The annual risk tends to be on the higher side, about 0.5–1% per year (1) for aneurysms that are 7–10 mm or larger in size; (2) for posterior circulation aneurysms; and (3) for aneurysms in patients with multiple brain aneurysms who have already ruptured another brain aneurysm [23]. Coiling can be an effective treatment option for unruptured aneurysms [24], and the best case for treatment might be an unruptured aneurysm that meets one of these three criteria and is amenable to coiling. The indications for open surgery for unruptured aneurysms are unclear but, if indicated at all, would likely be for one of the above scenarios in which coiling was not possible. Of note, the risks of serious complications from craniotomy for clipping an unruptured brain aneurysm are not insignificant, and likely in the range of 15–17% [22].

The rare unruptured aneurysm that presents with symptoms needs separate consideration. One special case is that of a posterior communicating artery (PCOM) aneurysm that presents with an acute third nerve palsy. Such an aneurysm should be promptly coiled (or clipped, if coiling is not possible) to avert an impending rupture. Also, giant aneurysms that present with symptoms due to thrombosis might be candidates for endovascular treatments.

The benefits for open surgery for unruptured aneurysms are often greatly overestimated, and the complication rate for such surgery is greatly underestimated.

Brain Arteriovenous Malformations and Arteriovenous Fistulae (AVM/AVF): Ruptured

These vascular anomalies are thought to usually be congenital, with a small percent being acquired lesions. If these present with a hemorrhage in a viable patient, they should be treated if possible. Very small brain AVMs and brain AVFs that can be "embolized for cure"—treated so as to completely eliminate the nidus or fistula—should be embolized. Also, if there is an associated aneurysm that has ruptured, then prompt embolization or coiling of that aneurysm would be appropriate. AVMs that cannot be completely embolized, and that are not extremely large, can be effectively treated with appropriate radiosurgery equipment, such as Gamma Knife [25] (see Figs. 5.1 and 5.2). For Gamma Knife, the preferred dose is 20–24 Gy to the 50% isodose line, depending on the size and location of the AVM/AVF. Certain larger lesions can sometimes be treated with radiosurgery in stages. (Radiosurgery results are better when the lesion is not embolized first.) Also, the AVM nidus is often not as large as people think and may well be amenable to radiosurgery treatment.

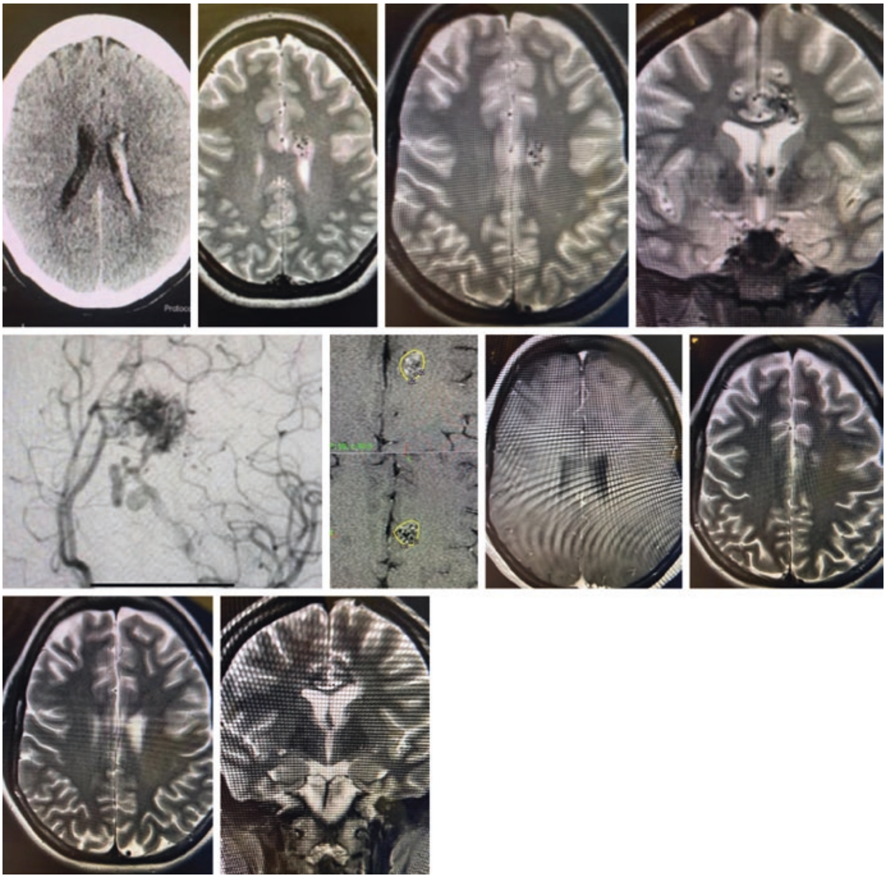

Fig. 5.1 This is a 25-year-old woman who presented with a sudden severe headache found to have an acute left intraventricular hemorrhage (**a**: axial CT image). MRI demonstrated a deep left frontal AVM extending into the left lateral ventricle (**b**, **c**: axial T2 weighted MRI; **d**: coronal T2 weighted MRI; **e**: lateral cerebral left carotid angiogram). The patient fully recovered from the bleed and subsequently underwent Gamma Knife treatment (**f**: axial T1 postcontrast images from the day of Gamma Knife treatment). MRI images 1.5 years after treatment show the AVM is gone (**g**: T1 postcontrast axial MRI; **h**, **i**: T2 weighted axial MRI; **j**: coronal T2 weighted MRI)

Certain lesions, however, are so large that no treatment can reasonably cure the abnormality. If the lesion cannot be completely embolized, and the patient is young and healthy, and the lesion is small and superficial, in non-eloquent cortex, and can be mostly embolized, then open surgery can be considered. It should be rare that a patient with a ruptured AVM or AVF is offered open surgery, over embolization, radiosurgery, or observation only. The complication rates for surgical removal are much higher than are generally appreciated.

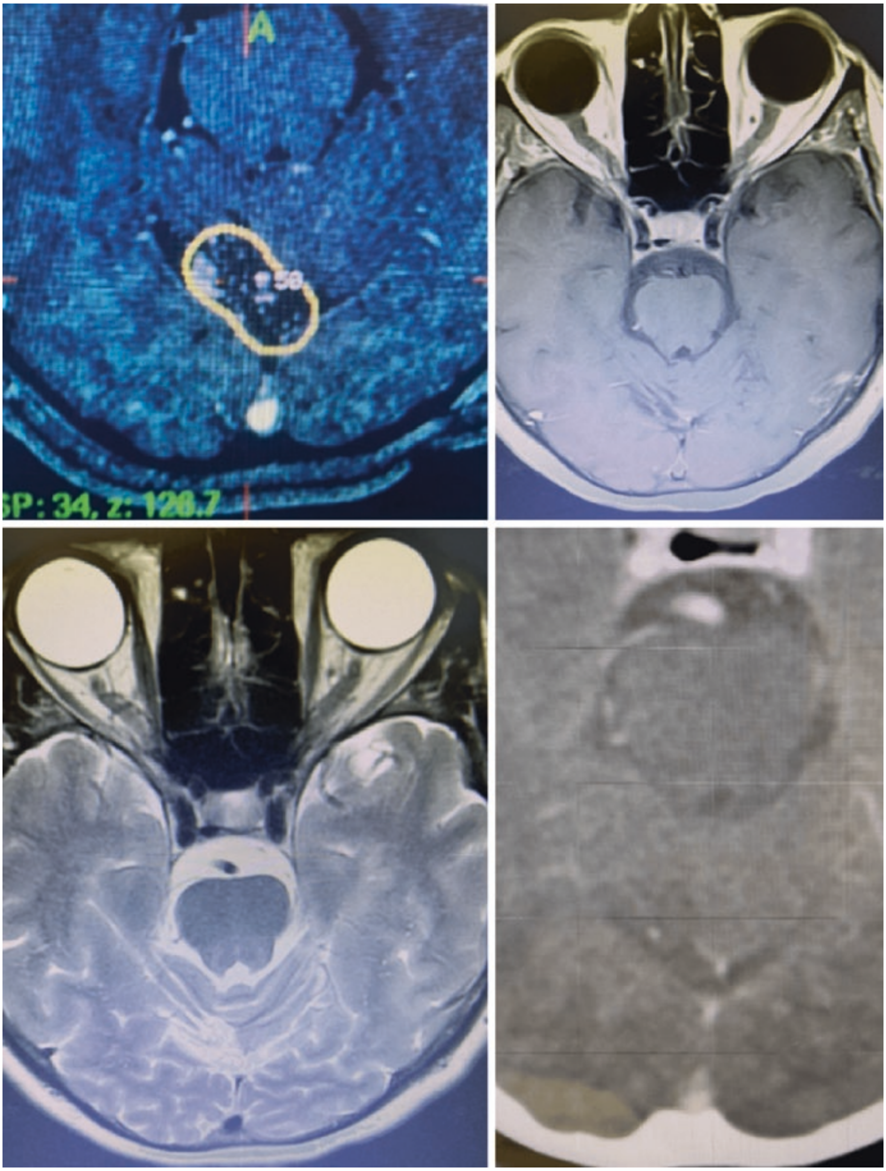

Fig. 5.2 This is a 40-year-old woman who presented with a severe headache and a small bleed from a posterior fossa AVM (upper cerebellar vermis) in her fifth month of pregnancy. She fully recovered and delivered a normal baby. Subsequently, she underwent Gamma Knife treatment (**a**: axial T1 postcontrast images on the day of treatment). She had no subsequent problems, and her follow-up images were showing that the AVM was starting to resolve. She was then lost to follow-up and returned 19 years later to get a check up on things. Follow-up images (**b**: axial T1 postcontrast MRI; **c**: axial T2 weighted MRI; **d**: axial CTA) showed complete resolution of the posterior fossa AVM

Brain AVM/AVF: Unruptured

An unruptured brain AVM/AVF is at lower risk of hemorrhage than a ruptured AVM/AVF. The ARUBA trial [26] looked at patients with unruptured brain AVMs that were thought to be “treatable” and compared those patients who were treated in any manner to those who were not treated. The study showed that the complication rate in the treated group was so high—and so much higher than what surgeons themselves usually reported—that they were expected to never be justified in light of the natural history. Another study showed similar results [27]. However, these studies did not substratify the treatment groups and did not look at, for example, Gamma Knife treatment only versus “no treatment.” As such—given the extremely safe profile for Gamma Knife and the real risk of these lesions—treatment with Gamma Knife is likely the best option (see Figs. 5.3 and 5.4). For AVM/AVFs that are tiny that can be cured with embolization alone, this may be performed instead. For AVM/AVFs that are very large, observation only is appropriate. Craniotomy is not clearly helpful. Again, if there were a case in which it might help, it would be for a young, healthy patient, with a very small AVM/AVF, on the surface, in a non-eloquent region, that could be mostly embolized prior to surgery.

Cavernous Malformations: Symptomatic

Most often, when cavernous malformations do cause symptoms, it is because of a small hemorrhage, particularly in a lesion that occurs in an eloquent region. Less commonly, these can present with seizures.

As most symptomatic brain cavernous malformations occur in deep and eloquent locations, an appropriate first line treatment option is usually Gamma Knife [28]. It is rare that patients will require craniotomy. Due to the location of these lesions, surgery is usually high risk and is not clearly better than the alternatives.

Cavernous Malformations: Asymptomatic

These should almost always be left alone. One could, however, argue that for a cavernous malformation in or by an eloquent region, clearly enlarging over time, Gamma Knife would be an appropriate treatment.

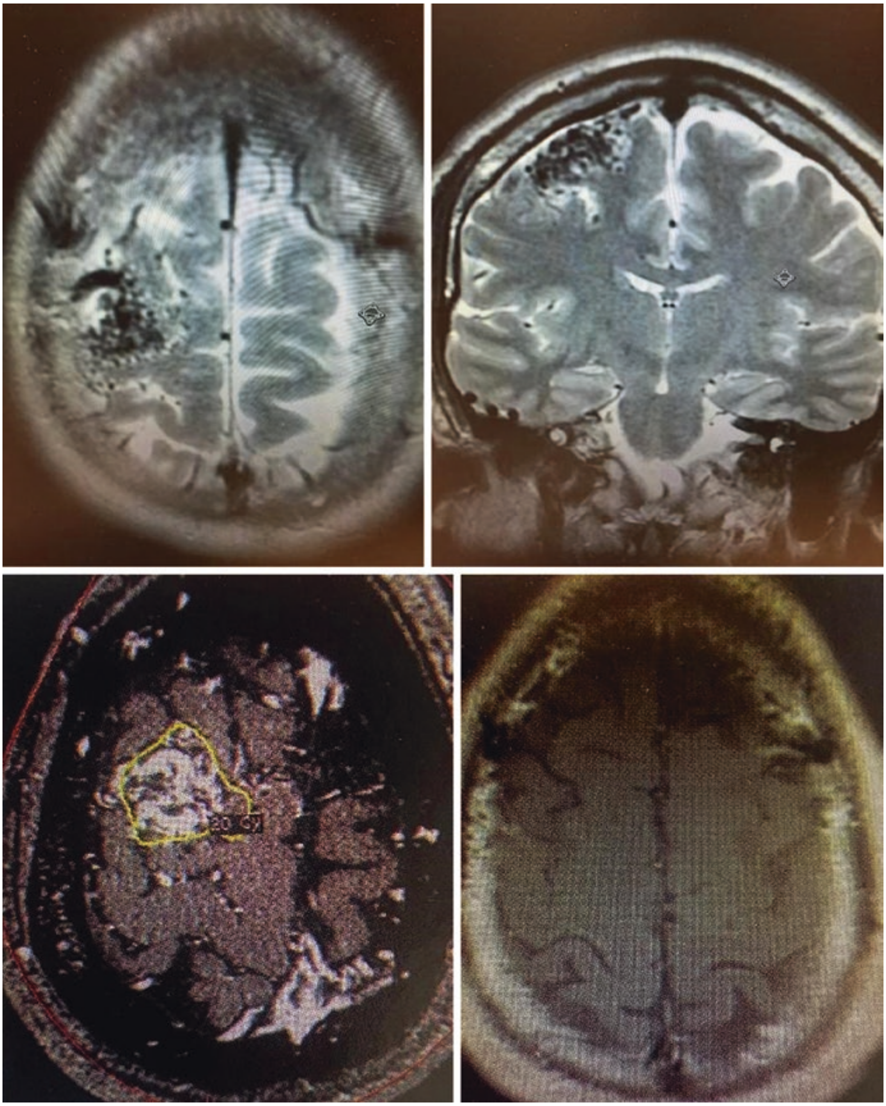

Fig. 5.3 This is a 28-year-old man who presented with headaches and was found to have a 2.5 cm right frontal AVM (**a**: axial T2 weighted MRI; **b**: coronal T2 weighted MRI). Gamma Knife was performed (**c**: day of Gamma Knife treatment, postcontrast T1 axial image). Three years later the AVM is gone (**d**) and he has no symptoms

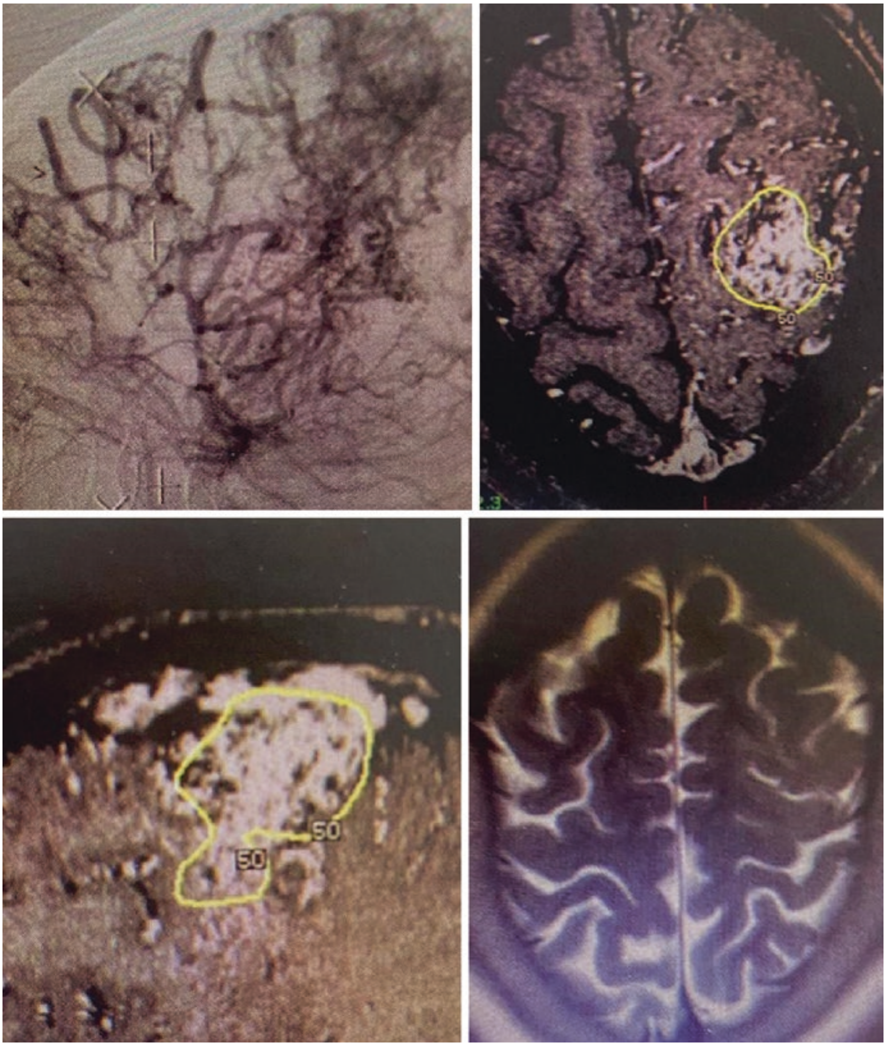

Fig. 5.4 This is a 13-year-old girl who was experiencing right arm and leg tingling episodes and headaches, found to have a moderate sized left frontal AVM with partial moyamoya disease (**a**: the AVM and moyamoya on lateral left carotid angiogram). Gamma Knife was performed (**b**: axial postcontrast T1 MRI from date of treatments; **c**: sagittal postcontrast T1 MRI from date of treatment). MRI from 6 years later shows complete resolution of the AVM (**d**: axial postcontrast T1 weighted MRI). Her symptoms have all resolved

Stroke

Stroke, with Large Vessel Occlusion

There is good evidence that viable patients with acute large vessel occlusions will, on average, benefit from prompt endovascular thrombectomy [29, 30].

Middle Cerebral Artery (MCA) Stroke

Numerous studies—The DESTINY trial [31], the DECIMAL trial [32], the HAMLET trial [33–35], the HeADFIRST trial [36], the DESTINY 2 trial [37], the HeMMI trial [38]—all showed no significant increase in the percent of patients who survived with good functional outcomes after hemicraniectomy for MCA stroke (hemicraniectomy did however significantly increase the percent of survivors with poor functional outcomes). Hemicraniectomy patients who survived were about five times more likely to have a lifelong dependence on others for care (Modified Rankin Scale/MRS 3, 4, or 5) than they were to be functionally independent (MRS 2). No patients survived with an MRS of 0 or 1.

While aggregating data from some of these studies suggested the possibility of a benefit, meta-analysis data found the quality of evidence that hemicraniectomy improved the likelihood of a good functional outcome in patients with MCA strokes to be "low" [39]. Even this conclusion required consideration of (1) studies that themselves showed no benefit; (2) studies that were stopped early; (3) studies that had very different inclusion requirements and different treatment methods; (4) studies that lumped together dominant and non-dominant MCA strokes, partial and complete distribution MCA strokes, and MCA strokes that were isolated as well as MCA strokes concurrent with strokes in other vascular distributions; (5) studies that included patients with significant disability who required lifelong assistance (MRS 3 and 4) as "good functional outcomes." Of interest, the DESTINY 2 trial, of patients over 60 treated with or without hemicraniectomy after a moderate to large MCA stroke, found that not one patient survived who was functionally independent (MRS 0, 1, or 2), and 80% of surviving patients, both surgically and medically managed, subsequently suffered from severe depression.

Nonetheless, there may be some very select group of young healthy patients who suffer acute moderate to large right-sided (non-dominant hemisphere) MCA strokes, and who would otherwise herniate, who may benefit from hemicraniectomy, if the patient (or family) is willing to accept that moderate to severe disability is a much more likely outcome than functional independence. This may be one of the few reasons to even consider a hemicraniectomy. Craniectomy in these cases, for optimum decompression, should be large, and the dura should be widely opened (but can then be covered with a large piece of allograft dura). As benefit is very uncertain for surgery even for an isolated right-sided MCA stroke, there would seem to be little justification for such surgery in the face of a left MCA stroke or an MCA stroke with concurrent strokes in other distributions, as survivors will be left with significant disability. A study that looked at hemicraniectomy for otherwise healthy patients under 60 with isolated right MCA strokes would not be unreasonable.

Cerebellar Stroke

Patients with large cerebellar stroke have a much poorer outcome if there is concurrent infarct of the brainstem or contralateral cerebellum. For large unilateral cerebellar stroke in otherwise viable patients, treatment of hydrocephalus with a ventriculostomy significantly increases the percent of patients with good functional outcomes. Performance of suboccipital craniectomy—whether with duraplasty, removal of infarcted cerebellum, or with ventriculostomy—does not clearly increase the percent of patients who survive with good functional outcomes compared with ventriculostomy alone [40–42]. That being said, there may be a select subgroup of younger, healthier, otherwise viable patients with large unilateral cerebellar strokes (in the absence of brainstem stroke or contralateral cerebellar stroke) who may benefit from suboccipital decompression in addition to ventriculostomy, and this would be worth further investigation.