Cerebral AVMs and Dural AVFs: Pathology and Management

The first of a three-part series on diagnosing, treating, and researching management options for intracranial malformations.

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Therapeutic embolization is a mainstay in neurosurgical management of a variety of pathologies, including intracranial malformations. In this three-part series, we start by introducing this relatively prevalent pathology with a particular focus on cerebral arteriovenous malformations (AVMs) and dural arteriovenous fistulas (DAVFs). The next article in the series will present current treatment options for cerebrovascular lesions while highlighting limitations of current clinical techniques with a focus on materials used in practice. The final article in the series will summarize ongoing research and future directions for the development of novel therapeutics.

REVIEW OF VASCULAR MALFORMATIONS

Embolization of vascular malformations is often indicated as an adjunct to surgery or radiosurgery and can be curative.[1] Therapeutic usage of endovascular embolization is most commonly indicated for occlusion of AVMs, DAVF tumor vessels, and aneurysms.[2-5] AVMs are defects in the circulatory system, characterized by abnormal communications between arteries and veins and are commonly congenital (Figure 1A–1C). Treatment of intracranial tumors can also be achieved through endovascular embolization of feeder vessels. This technique is often indicated in tumors with a discernible vascular mass that can be embolized to minimize blood loss and damage to normal tissue. It also further diminishes the angiogenic response and complements conventional adjuvant, neoadjuvant, or surgical tumor therapy options.

Although the focus of this review is on AVMs and DAVFs, aneurysms are another type of vascular anomaly that can be treated through endovascular embolization, which is frequently discussed as fusiform aneurysm filling and saccular aneurysm filling. Fusiform aneurysms are a diffuse dilatation of vasculature. In contrast, saccular aneurysms are localized outpouchings or sac-like protrusions of arterial vessels (Figure 1A). These aneurysms are typically found at vessel bifurcations and are the most common type of intracranial aneurysm and cause of aneurysmal subarachnoid hemorrhage. Due to their potentially devastating consequences, saccular aneurysms are carefully followed and treated through either surgical clipping or endovascular repair (eg, coils, stents, and flow diverters).

Though the uses of liquid embolics in the cerebral vasculature are numerous, the remainder of this review will focus on the different materials used to occlude AVMs and DAVFs.

PATHOLOGY AND PREVALENCE

Cerebral AVMs are abnormal connections between arteries and veins in the brain and occur in < 1% of the population, with approximately 12% of cases being symptomatic.[6] Most commonly found in adults aged 20 to 40 years, cerebral AVMs hemorrhage at a rate of about 2% to 3% per year, half of which carry significant morbidity and mortality rates.[6,7] Although intracerebral hemorrhage occurs more commonly and is life-threatening, subarachnoid hemorrhage (SAH) may also result in lifelong complications and disabilities due to an increase in pressure on the brain. Depending on the location, intracerebral hemorrhage can lead to SAH if the bleeding

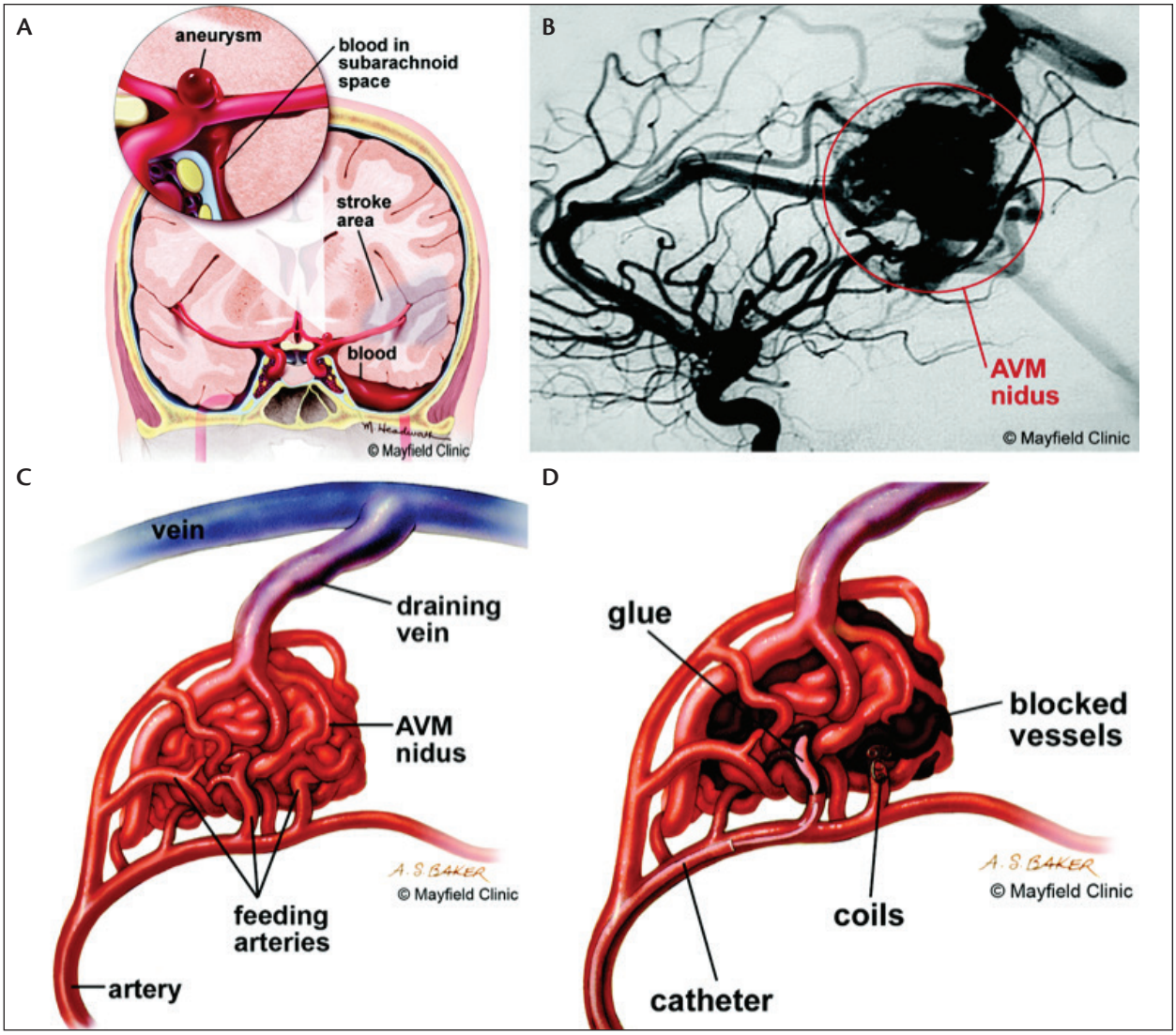

Figure 1. Pathology of cerebrovascular malformations. A cascade of hemodynamic, metabolic, and cellular factors affect development of aneurysms (A). Arteriovenous malformations can be diagnosed using CT or MRI with subsequent cerebral angiography, such as the one seen in the parietal lobe, which may lead to intracerebral hemorrhage (ICH) if untreated (B). Schematic of the pathology (C) and potential treatment options (D). Reprinted with permission from MayfieldClinic.com.

dissects either through the surface of the brain or into the intraventricular space. Seizures are the second most common presentation in AVMs, occurring in approximately 20% of cases, followed by headaches (15%), focal neurological deficit (< 5%), and pulsatile tinnitus.[6] Similarly, dural arteriovenous fistulas (DAVFs) are abnormal connections between arteries and veins or venous spaces called *sinuses*. AVMs are found within the brain parenchyma or spinal cord; DAVFs are found in the dura mater or arachnoid covering the brain or spinal cord. The feared consequence of these abnormal connections is the transfer of high-pressure flow from the arteries to cortical veins, which can potentially cause irreversible damage to the brain or spinal cord via hemorrhage or venous congestion.[6,8]

Individuals with cerebral AVMs are at a moderately high risk for hemorrhage and seizures, depending on size and location of the AVM and patient demographics.[9] Patients with AVM-related hemorrhages have traditionally undergone invasive treatment to prevent subsequent hemorrhage. With the advent of contemporary brain imaging techniques, patients diagnosed with unruptured cerebral AVMs are now considered candidates for treatment. Treatment planning for AVMs and DAVFs has

evolved substantially to become less invasive and more effective at eliminating the vascular lesions.[10,11] A large, multicenter, international clinical study looking into the benefits of medical management with and without intervention (the ARUBA trial) demonstrated that interventional therapy may in fact increase chances of symptomatic stroke and mortality for unruptured cerebral AVMs.[12] However, this study, while recent, reports on 33-month data, and longer-term 5-year clinical evaluation is yet to be reported.[12]

Current treatment options to reduce the risk of hemorrhage or facilitate subsequent surgical removal of an AVM or DAVF include endovascular injection of occlusive agents or radiosurgery.[6,13] Although a variety of embolic agents (solid and liquid, permanent and temporary) have been developed and implemented in humans, the need for less invasive, more biocompatible, and cost-effective methods still exists. Though the durability of endovascular treatment alone is not certain, it is a useful adjunct before surgery to reduce blood loss and operative times, hence improving outcomes. Endovascular embolization may offer a curative role in well-selected cases with a small nidus and low numbers of arterial feeders.

Evaluation of patient history and epidemiology of potential treatment strategies and outcomes for intracranial AVMs has led to development of improved treatment modalities.[6,7,14] Because AVMs represent a relatively rare and heterogenous group of cerebrovascular lesions exhibiting different natural histories, there are inherent challenges when comparing the efficacy of various treatment methods. From the study of the natural history of this pathology, we know that the mortality rate from the first hemorrhage is between 10% and 30%, with 10% to 20% of AVM-related hemorrhage survivors experiencing some form of long-term disability.[6] Possible predictors of hemorrhage such as seizures, AVM size, feeding artery pressures, characteristics of the venous drainage system, and AVM location have not been observed consistently across previous studies.[6] Thus, the complex natural history of these cerebrovascular malformations provides challenges to treatment planning.

CONCLUSION

In this series, we hope to give the readership a view into neurosurgical management of cerebrovascular pathologies with a focus on vascular malformations. Cerebral AVMs and DAVFs are relatively prevalent in the population, with major concerns of hemorrhage and intracranial bleeding if left untreated. Ultimately, patient history/candidacy, surgeon adeptness, and available technologies influence treatment plans. The first part in this series has helped identify the pathology and set the stage for the next installment detailing management of these pathologies. ■

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