Cerebral Angiography: Arteriovenous Malformations and Dural Arteriovenous Fistulae

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Pathophysiology

Arteriovenous Malformations (AVM)

Intracranial arteriovenous malformations (AVMs) are abnormal communications between pial arteries and veins with an intervening abnormal tangle of vessels termed a "nidus." The abnormal vessels are prone to bleeding and rupture. The lack of normal intervening capillaries increases pressure in the draining veins, which further potentiates the risk of rupture. The cause of intracranial AVMs is unknown but is thought to be either secondary to abnormal intrauterine vascular development or as a response to a prior vascular insult [1]. Symptomatic AVMs are estimated to occur in approximately 1 in 100,000 people [2]. Many AVMs occur sporadically; however some familial syndromes predispose to their development, i.e., hereditary hemorrhagic telangiectasia. Presenting symptoms are often nonspecific and include headaches, seizures, and intracranial hemorrhage (ICH). Certain AVM features are associated with an increased risk of rupture including prior hemorrhage, deep venous drainage, and perinidal or intranidal aneurysm [3]. AVMs are classified utilizing the Spetzler-Martin grading system, which stratifies lesions based on surgical risk (Table 47.1). Larger lesions, those with drainage to the deep venous system and those which occur in eloquent areas of the brain, are at increased risk of surgical complications during resection [4].

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Dural Arteriovenous Fistulas (DAVF)

Dural arteriovenous fistulas (DAVF) are abnormal communications between meningeal arteries and dural venous sinuses. DAVF account for approximately 10–15% of intracranial vascular malformations. DAVF can be idiopathic, posttraumatic, and postsurgical, or they can occur following dural venous thrombosis in hypercoagulable individuals. Patients can present with very specific clinical symptoms, which can often help localize the lesion, including unilateral pulsatile tinnitus or unilateral scleral injection, eye pain, proptosis, or visual changes [5]. In some cases, symptoms can be very nonspecific such as altered mental status, headaches, or ICH. "Benign" DAVF only result in irritating symptoms for the patient, whereas "malignant" DAVF place the patient at a very high risk of ICH. There are two main grading systems of DAVF, the Borden classification system and the Cognard classification system (Table 47.2) [6, 7]. DAVF grade is predominantly determined by the venous drainage pattern. Lesions with retrograde or direct flow into cortical veins place patients at higher risk of ICH. If these draining cortical veins are ectatic (dilated), patients are at an even greater risk of intracranial hemorrhage [8, 9]. AVM and DAVF are compared in Table 47.3.

Key Point

Intracranial AVM = communication between pial arteries and veins with intervening nidus.
Dural AVF = communication between meningeal arteries and dural venous sinuses

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Table 47.1 Spetzler-Martin grading classification: AVMs

Size of nidus	Small (<3 cm) = 1
	Medium (3–6 cm) = 2
	Large (>6 cm) = 3
Eloquence of adjacent brain	Non-eloquent = 0
	Eloquent = 1
Venous drainage	Superficial only = 0
	Deep = 1

The three characteristics of the AVM are added together to give a grade between 1 and 5. The score correlates with the operative outcome with a higher score corresponding with an increased risk of a poor outcome

Table 47.2 Classification systems for DAVFs

	High-grade DAVF	Low-grade DAVF
Borden classification	2, 3	1
Cognard classification	IIb, IIa + b, III, IV	I, IIa
Venous drainage characteristics	Cortical venous reflux Direct cortical venous drainage Cortical venous ectasia	Antegrade drainage through a dural sinus or meningeal veins
Common symptoms	Headache, AMS, ICH	Tinnitus, visual changes, proptosis, eye redness

AMS altered mental status ICH intracranial hemorrhage

Table 47.3 Comparison of cerebral arteriovenous malformation (AVM) and dural arteriovenous fistula (DAVF)

	AVM	DAVF
Abnormal vessels	Pial arteries and veins	Meningeal arteries and dural venous sinuses
Cause	Intrauterine vascular development Prior vascular insult Syndromic (HHT)	Posttraumatic Postsurgical Secondary to dural venous thrombosis Idiopathic
Symptoms	Nonspecific: headaches, seizure, ICH	Localized: unilateral pulsatile tinnitus, etc. Nonspecific: AMS, headache, ICH
Classification system	Spetzler-Martin system (based on surgical risk)	Borden and Cognard systems (based on venous drainage pattern)

HHT hereditary hemorrhagic telangiectasia, ICH intracranial hemorrhage. AMS altered mental status

Clinical Indication

AVM

AVMs are often found incidentally on cerebral imaging or found when the patient presents with seizure or ICH. While a variety of neurologic exam findings can be seen with ruptured AVM, unruptured AVMs are often asymptomatic. CTA and MRI/MRA can show hypertrophied feeding arteries and draining veins and a characteristic "bag of worms" appearance [10, 11]. MRI can also show prominent flow voids in the area of the lesion. Catheter-based digital subtraction angiography (DSA) is the gold standard technique for the evaluation of AVM because it allows for intricate assessment of the AVM nidus, identification of all feeding arteries and draining veins, and it is sensitive for the detection of any related aneurysms (Fig. 47.1). Indications for treatment include prior rupture/ICH, significant clinical symptoms, and, in some cases, coexistent aneurysms [12]. Treatment of symptomatic lesions (those with prior hemorrhage) is indicated to prevent rerupture. Treatment of asymptomatic lesions remains controversial and varies widely by institution.

DAVF

The diagnosis of DAVF commonly occurs when patients present with new onset of neurologic symptoms. Physical exam findings may be generalized such as headache or localized; localized symptoms can often elucidate the location of the lesion. Pulsatile tinnitus or diminished hearing may indicate a fistula near the internal auditory canal. A caroticocavernous fistula may produce visual symptoms such as proptosis, conjunctival injection, and optic disc herniation [13]. Patients with malignant lesions may demonstrate symptoms related to ICH including altered mental status, somnolence, asymmetric pupillary dilation, and papilledema [14]. CT and MRI can show malignant features such as dural venous sinus thrombosis, venous infarct, ICH, brain parenchymal edema, and hypertrophied external carotid branches [5] (Fig. 47.2). The gold standard imaging test remains catheter-based DSA as it can define the fistulous point, dynamically evaluate flow patterns, and assess for any cortical venous drainage. Indications for treatment include ICH, malignant features on imaging, and intractable debilitating symptoms.

Key Point
The grade of DAVF depends predominantly on venous drainage.

Conventional Therapy

AVM

Surgery remains the first-line treatment option in surgically accessible lesions as it has a success rate approaching 100% and has acceptable complication rates [15, 16]. During surgery, arterial feeding vessels and draining veins are identified. Arterial feeders are disconnected from the AVM nidus using coagulation and surgical ligation. Following disconnection

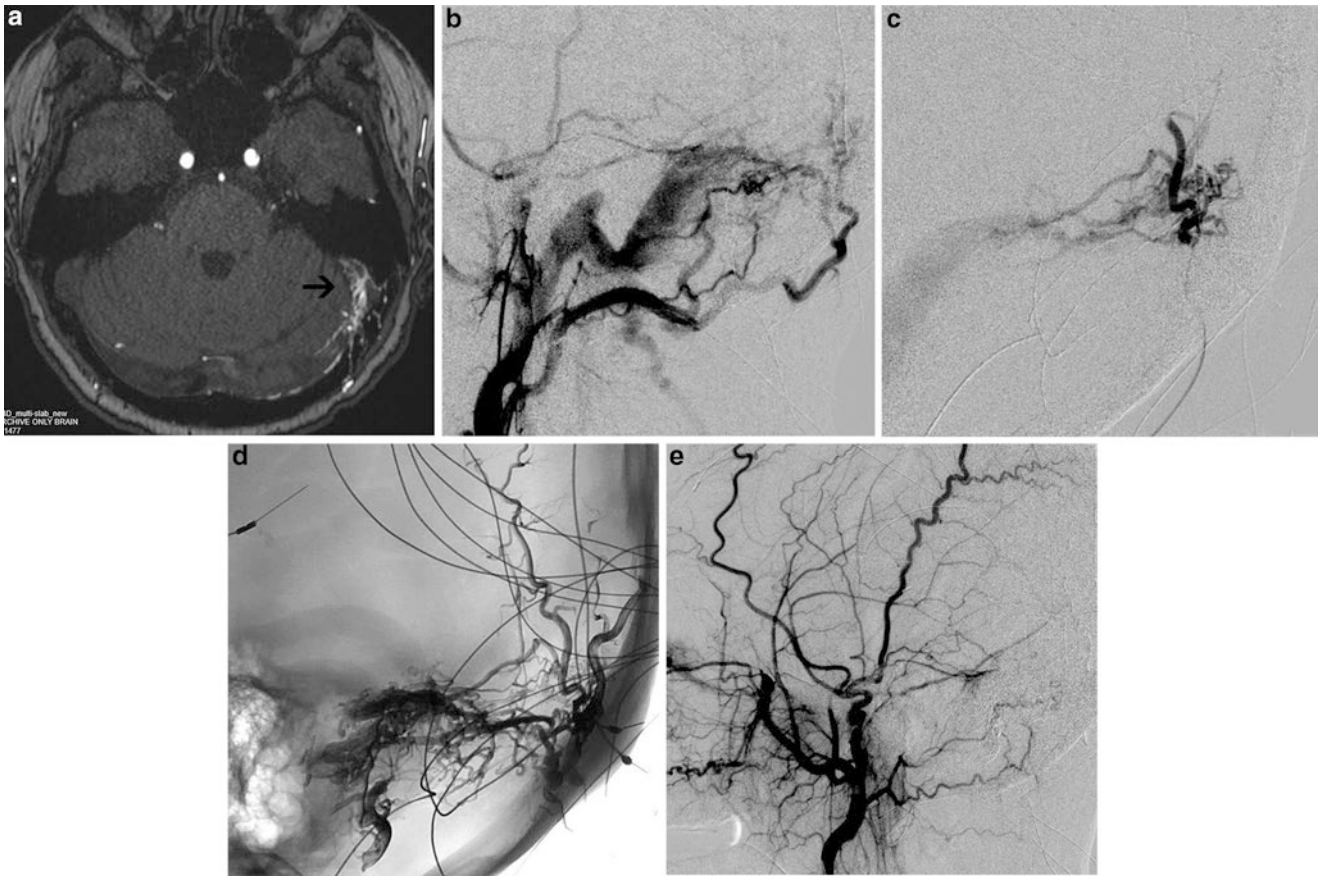

Fig. 47.1 A 55-year-old female who presented with tinnitus and headache. (**a**) Axial time of flight (TOF) MRA source image shows multiple arterial/venous fistulae in the left transverse and sigmoid sinus (black arrow). (**b**) Lateral left external carotid angiogram (LECA) shows a dural arterial venous fistula (DAVF) with multiple arterial branches from the left occipital artery, middle meningeal artery, and posterior auricular artery filling the sigmoid sinus. (**c**) Lateral selective angiogram of the transmastoid branch of the left occipital artery shows multiple arterial venous fistulae draining into the sigmoid sinus. (**d**) Lateral spot fluoroscopic image shows the Onyx cast within the DAVF following embolization. (**e**) Lateral left external carotid angiogram after embolization with Onyx shows no filling of the DAVF

of arterial feeders, the draining veins are resected near the end of excision. Stereotactic radiosurgery (SRS) remains the treatment of choice if the AVM is unresectable [12]. In some cases, combination therapy with open surgery, endovascular intervention, and/or SRS can be used.

DAVF

Benign DAVF are often managed expectantly. In some cases, spontaneous regression may occur [17]. Intermittent manual compression of the ipsilateral carotid artery can also be performed, as this can lead to fistulous occlusion [18]. Rarely, benign DAVF can progress to malignant lesions, but this is usually accompanied by a change in symptoms [19]. In the past, surgery has been the treatment of choice for malignant DAVF, by aiming to surgically transect the abnormal communication by ligating the fistulous communication. With advances in technology, endovascular therapy has surpassed surgery as the standard treatment for DAVF.

Interventional Therapy

AVM and DAVF

Endovascular therapy for AVM is usually used as an adjunct to surgery or SRS [20]. Endovascular therapy is often directed at occluding specific feeding vessels or portions of the AVM which are not accessible to the operative surgeon or SRS. Additionally, endovascular therapy can be used to target perinidal aneurysms which are at increased risk of rupture [21]. Endovascular AVM treatment usually employs liquid embolic agents which penetrate and occlude the AVM nidus.

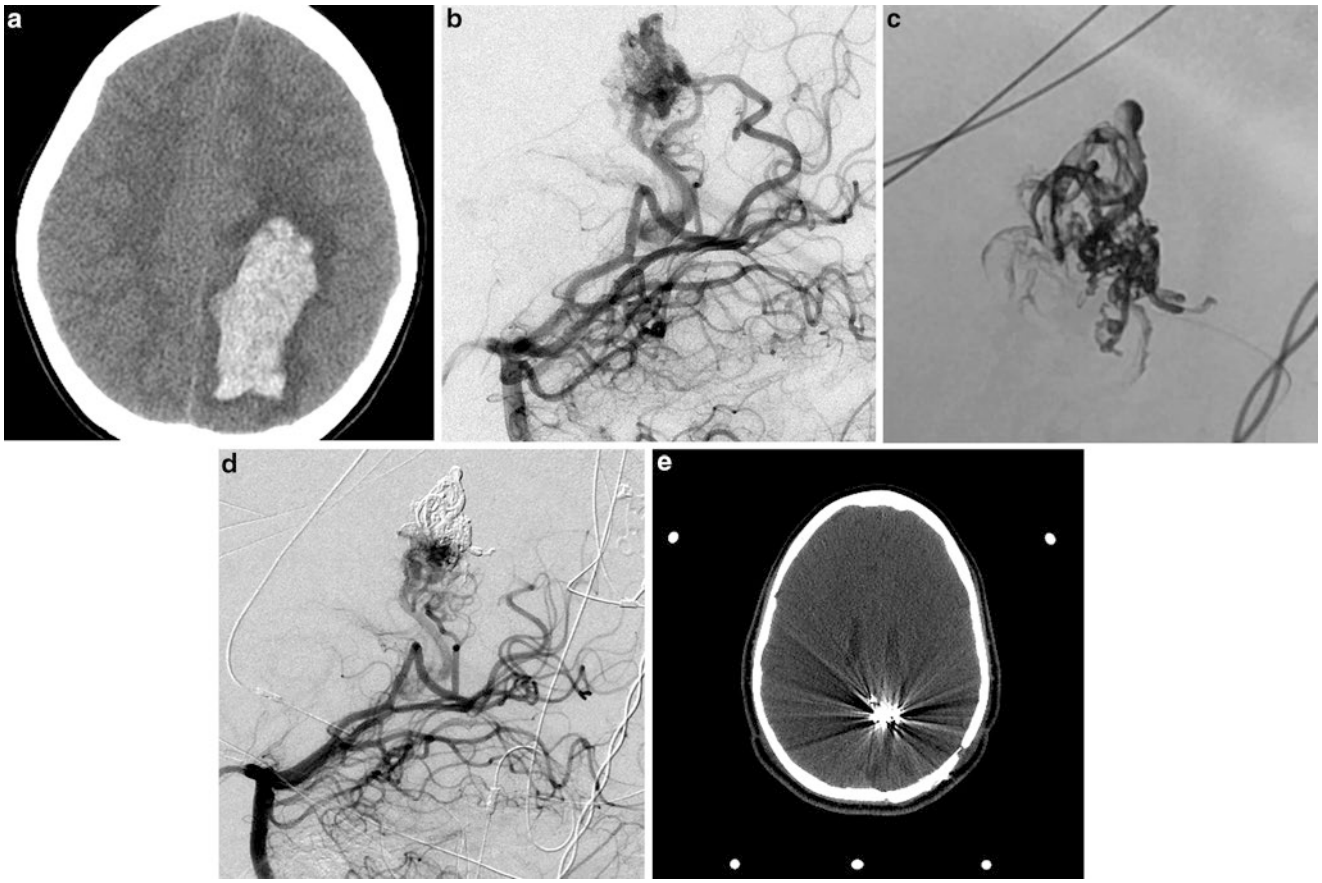

Fig. 47.2 An 11-year-old female who presents with acute onset of right arm and leg weakness with sensory changes. (**a**) Axial non-contrast head CT shows an intraparenchymal hemorrhage in the left motor and sensory regions of the brain. (**b**) Lateral left vertebral angiogram (LVA) shows an AVM nidus in the area of prior hemorrhage identified on the head CT. There is a small intranidal aneurysm, which is the assumed area of hemorrhage. (**c**) Lateral spot fluoroscopic image shows the Onyx cast in the AVM and intranidal aneurysm following embolization. (**d**) Lateral LVA post-embolization shows occlusion of the portion of the AVM nidus embolized with Onyx. (**e**) Axial CT scan of the head for stereotactic radiation. Note the Onyx cast causing streak artifact

For DAVF, endovascular treatment is often the best treatment option. Treatment of DAVF can be achieved via arterial or venous approaches. Venous approaches aim to occlude the venous pouch at the fistulous point with coils. Arterial approaches utilize liquid embolic agents to penetrate and occlude the fistulous communication. Advances in endovascular technology have allowed for high success rates and low complication rates in the treatment of DAVF [22].

AVM and DAVF should be evaluated with catheter-based DSA prior to treatment to fully delineate arterial feeders and venous flow patterns. Often, cases are presented at multidisciplinary conference to obtain multi-specialty consensus on the best treatment approach. Standard preoperative labs for arterial intervention should be performed including CBC, coagulation labs, and a BMP. Procedural risks should be discussed with the patient and/or health care provider and should include non-target embolization resulting in stroke, intracranial vessel injury resulting in ICH, and lesional rupture resulting in ICH. Cranial nerve injuries should be a focus in DAVF, as meningeal vasculature often contributes to cranial nerve blood supply.

Key Point

Complications of AVM and DAVF treatment:

- Stroke from nontarget embolization
- ICH from vessel injury or lesion rupture
- Cranial nerve injury for DAVF treatment

The How To

1. Femoral Artery access is obtained utilizing ultrasound and fluoroscopic guidance via the Seldinger technique. If venous angiography or intervention is planned, a venous sheath should also be placed.
2. Patients without acute rupture are therapeutically heparinized to 2–2.5 times baseline activated clotting time (ACT) (generally 50–100 units/kg). Therapeutic heparinization is maintained throughout the procedure.
3. Usually, procedures begin with a six vessel diagnostic cerebral angiographies to fully evaluate contributions to the lesion.
4. Following diagnostic angiography, the diagnostic catheter is exchanged over a wire for an interventional guide catheter. Utilizing a negative road map, a microcatheter and microwire are advanced through the guidecatheter as close as possible to the level of the AVM nidus or fistulous point (see Fig. 47.2).
5. In lesions supplied by long tortuous vessels, additional stability can be obtained by advancing an intermediate catheter through the guide catheter, followed by the microcatheter and microwire.
6. Liquid embolic materials have become the agents of choice in most cases of DAVF and AVM treatment (see Fig. 47.1). Ethylene vinyl copolymer (EVOH) is a viscous cohesive agent which allows for a slow, controlled injection [23]. Additionally, EVOH contains tantalum powder, which allows for easy visualization under fluoroscopy. N-butyl cyanoacrylate (NBCA) is a low-viscosity adhesive liquid embolic agent which rapidly polymerizes when it contacts blood [23]. The rapid polymerization can make embolization with NBCA more technically challenging.
7. Regardless of embolic agent, the goal of AVM treatment is penetration and occlusion of the nidus, and the goal of DAVF treatment is occlusion of the fistulous point with cure of all malignant features.
8. Post-procedural angiography should be performed to assess for any residual lesional flow and to act as a baseline for follow-up studies.

Post-procedurally, patients should be monitored in a neurologic ICU for at least 1 day.

Key Point
Diagnostic cerebral angiogram consists of visualizing six vessels:

- Bilateral internal carotid arteries
- Bilateral external carotid arteries
- Bilateral vertebral arteries

Key Point
Treatment goals:

AVM = penetration and occlusion of the nidus.
DAVF = occlusion of the fistulous point with treatment of all malignant features

Key Point
Lesions with cortical venous drainage must be treated until all cortical drainage is cured.

Imaging follow-up should be performed at regular intervals; timing is institutional specific but typically begins at 6 months post-treatment. Any recurrence or change in symptoms should prompt rapid reevaluation as this could indicate lesional progression or recurrence.

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