Endovascular Treatment of Cranial Dural Arterio-Venous Fistulas

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Abstract

Intracranial dural arteriovenous fistulas (DAVFs) are relatively rare, acquired, pathologic shunts between dural arteries and dural venous sinuses, meningeal veins, or cortical veins. Signs and symptoms are highly variable and depend upon several factors including fistula location, duration of disease, and venous drainage pattern. The most clinically important feature for classification is venous drainage pattern. Management decisions require a multidisciplinary approach involving discussions among interventional neuroradiologists, neurosurgeons, and neurologists and depend on the type of lesion, including location and angiographic risk strata, assessment of clinical presentation, and patient status (age, comorbidities). Treatment should be pursued for all lesions with cortical venous drainage or intolerable symptoms.

Glossary of Terms

Borden classification Classification system of dural arteriovenous fistulas where by these lesions are grouped into three types based upon the site of venous drainage and the presence or absence of cortical venous drainage.

Cognard classification Classification system of dural arteriovenous fistulas that correlates venous drainage patterns with increasingly aggressive neurological clinical course.

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Compression therapy Non-invasive method to treat cavernous sinus dural arteriovenous fistulae.

Dural arteriovenous fistulas An abnormal direct connection between a meningeal artery and a meningeal vein or dural venous sinus.

NBCA N-Butyl Cyanoacrylate is a liquid embolic used in the treatment of cerebral vascular malformations.

Onyx Non-adhesive liquid embolic agent used for the pre-surgical embolization of cerebral vascular malformations.

Introduction

Intracranial dural arteriovenous fistulas (DAVFs) are acquired, pathologic shunts between dural arteries and dural venous sinuses, meningeal veins, or cortical veins. The artery-to-vein connection is located within the leaflets of the dura mater and lacks a distinctive vascular nidus. By contrast, parenchymal arteriovenous malformations occur within the brain parenchyma and usually contain a vascular nidus. DAVFs are relatively rare accounting for 10–15 % of intracranial arteriovenous shunts and representing 6 % of supratentorial and 35 % of infratentorial vascular malformations (Newton and Cronqvist 1969; Al-Shahi et al. 2003; Chaudhary et al. 1982). Signs and symptoms are highly variable and depend upon several factors including fistula location, duration of disease, and venous drainage pattern. Symptoms range from pulse-synchronous tinnitus and exophthalmos to cranial nerve deficits, cognitive impairment, venous infarction, intracranial hemorrhage, and even death. DAVFs occur most commonly in the transverse/sigmoid and cavernous sinuses. The decision to treat, as well as the mode of treatment, should be based on the angiographic features of the lesion, including the venous drainage pattern, location of the DAVF, severity of presenting symptoms, the morbidity and mortality rate(s) of the procedure(s) under consideration, and general medical condition of the patient (Brown et al. 1994; Daniels et al. 2013; Natarajan et al. 2010; Hu et al. 2011).

Etiology/Pathophysiology

Although the pathogenesis of DAVFs is still a matter of debate, multiple associations have been described in the literature including head trauma, surgery, infection, hormonal factors, hypercoagulable states (e.g., protein S deficiency, prothrombin mutation, and factor V Leiden), and sinus thrombosis (Kawaguchi et al. 2002; Field et al. 2002; Hashimoto et al. 1998; Yoshimura et al. 1995; Chung et al. 2002; Nabors et al. 1987; Chaudhary et al. 1982; Nishio et al. 2002; Yassari et al. 2002; Witt et al. 1999; Terada et al. 1998; Singh et al. 2001). The association between sinus thrombosis and DAVFs has long been recognized, although the pathogenesis and evolution from thrombosis to fistula development are still not completely understood.

Two etiologic hypotheses regarding the association between sinus thrombosis and DAVFs have been proposed. The first theory suggests that venous hypertension and ischemia, resulting from sinus thrombosis/sinus outflow obstruction, induces the pathologic enlargement of physiologic but quiescent, arteriovenous shunts (between meningeal arteries and dural venous sinuses) within the dura mater to facilitate development of collateral blood flow (Nabors et al. 1987; Kerber and Newton 1973). These "microshunts" may continue to grow in patients with genetic predisposition or under the influence of other unknown factors and eventually develop into DAVFs. The second theory postulates that DAVFs are the result of vascular neogenesis within the dura mater resulting from the release of angiogenic growth factors. Elevated levels of both vascular endothelial growth factor and basic fibroblast growth factor have been found in the sinus wall and perivascular connective tissues of DAVFs in both humans and experimental rat models. These angiogenic factors can be either directly produced by inflammatory cells in organizing thrombus or indirectly induced by ischemia resulting from increased intraluminal venous pressure (Malek et al. 2000; Uranishi et al. 1999; Tirakotai et al. 2005; Lawton et al. 1997; Herman et al. 1995; Terada et al. 1994, 1996).

Classification

Over the last several decades, numerous classification schemes have been proposed in order to help define the natural history of DAVFs. Initial reports focused on the anatomical location as the key discriminating feature (Aminoff 1973). Although lesion location correlates well with symptomatology, it does not necessarily predict natural history, as DAVFs in any location can exhibit aggressive neurological behavior. It was soon recognized that the venous drainage pattern determines a DAVFs clinical behavior and was the most clinically important feature for classification (Brown et al. 1994; Awad et al. 1990; Castaigne et al. 1976; Houser et al. 1972).

Understanding this, Djindian and Merland developed a classification scheme dividing DAVFs into four types based on venous drainage pattern. Type I lesions demonstrate immediate drainage into a dural sinus or meningeal vein and are considered relatively benign. Type II lesions demonstrate initial drainage into a sinus and retrograde flow into other sinuses or cortical veins. Type III lesions demonstrate initial drainage into a cortical vein, and type IV lesions have drainage into a cortical vein with venous ectasia. Type II–IV lesions show increased risk of neurological symptoms and hemorrhage (Djindjian and Merland 1978).

Lalwani et al. proposed a system for grading DAVFs of the transverse/sigmoid sinus based on the severity of venous restrictive disease, as obstruction of venous outflow increases the risk of intracranial hemorrhage due to retrograde flow via cortical veins. Four grades of venous restrictive disease are defined. Critical factors include the presence or absence of cortical venous drainage and antegrade venous drainage through the ipsilateral sigmoid sinus and jugular bulb. Grade 1 lesions are characterized by normal antegrade venous drainage without venous restriction or cortical venous drainage. Grade 2 lesions demonstrate antegrade venous drainage and retrograde venous flow through the transverse sinus to the torcular Herophili, with or without cortical venous drainage. Grade 3 lesions have retrograde and cortical venous drainage without antegrade flow through the sigmoid sinus. Grade 4 disease is characterized by cortical venous drainage only, due to severe antegrade and retrograde venous obstruction in the transverse and sigmoid sinuses. Their preliminary study demonstrated that patients with grades 1 and 2 lesions typically had a benign clinical presentation, primarily pulsatile tinnitus and headache, while patients with grade 3 and grade 4 lesions had more ominous clinical presentations, typically visual symptoms and intracranial hemorrhage (Lalwani et al. 1993).

The Cognard classification system uses Djindian's scheme as a base but expands upon it to include five main types and several subtypes of venous drainage. Classifications are based on the direction of dural sinus drainage, the presence or absence of cortical venous drainage, and venous outflow architecture. Type I lesions drain into a main sinus and demonstrate antegrade flow. Type II lesions drain into a main sinus but demonstrate retrograde flow into either a main sinus (IIa), cortical veins (IIb), or both (IIa + b). Type III lesions show direct cortical venous drainage without venous ectasia. Type IV lesions have direct cortical venous drainage with venous ectasia. Type V lesions are intracranial DAVFs that drain into spinal perimedullary veins. In their series of 205 patients, only one of 84 patients with a type I DAVF had severe neurological symptoms; 83 patients had a benign course. In type II lesions, reflux into the sinus induced intracranial hypertension in 20 % of cases, and reflux into cortical veins induced hemorrhage in 10 %. Forty percent of patients with type III lesions had intracranial hemorrhage, and the presence of venous ectasia (type IV) increased the risk to 65 %. Type V lesions produced progressive myelopathy in 50 % of cases (Cognard et al. 1995).

The Borden classification system divides DAVFs into three types based on the site of venous drainage (dural sinus and/or cortical vein) and the presence or absence of cortical venous drainage. Type I lesions drain directly into dural venous sinuses or meningeal veins. Type II fistulas drain into dural sinuses or meningeal veins but also have retrograde drainage into subarachnoid veins. Type III lesions drain into

subarachnoid veins and do not have dural sinus or meningeal venous drainage. The Borden classification scheme further subclassifies DAVFs as either single-hole or multiple-hole lesions (Borden et al. 1995; Davies et al. 1996).

More recent classification schemes have been proposed that grade DAVFs through a combination of angiographic and clinical features. Zipfel et al. developed a classification scheme recognizing that DAVFs with high-risk angiographic features that present with intracerebral hemorrhage or nonhemorrhagic neurological deficits harbor more risk for future injury compared with DAVFs with high-risk features that have a benign clinical presentation. Their classification system divides DAVFs into three main types, with types 2 and 3 each having two subtypes depending on whether or not they present with symptomatic cortical venous drainage. A stepwise increase in neurological risk is noted between type 1 lesions and type 3 lesions (Zipfel et al. 2009).

The Borden and Cognard classification schemes, both of which highlight the importance of venous drainage patterns (particularly cortical venous drainage), are the two most widely used and clinically accepted classification schemes for DAVFs (Cognard et al. 1995; Borden et al. 1995; Davies et al. 1996; Table 1). In both classification schemes, the presence of cortical venous reflux is considered an aggressive feature denoting a high-risk lesion with an annual mortality rate of 10.4 % and an annual risk of hemorrhage and nonhemorrhagic neurological deficit of 8.1 % and 6.9 %, respectively (van Dijk et al. 2002). Regardless of the classification scheme one chooses to utilize, it is extremely important to be aware that the venous drainage patterns of DAVFs are dynamic and lower-grade lesions can progress to higher grades over time (Satomi et al. 2002; Cognard et al. 1997). Should a patient's symptoms change, reevaluation and follow-up imaging are warranted (Shah et al. 2012).

Fistulas involving the cavernous sinus have been traditionally divided into four types based on their arterial supply (Barrow et al. 1985). Type B DAVFs involve meningeal branches of the internal carotid artery; type C lesions are shunts between meningeal branches of the external carotid artery and the cavernous sinus; type D DAVFs are dural shunts between meningeal branches of both the internal and external carotid arteries and the cavernous sinus. Type A lesions, also called "direct" carotid-cavernous fistulas, are direct high-flow shunts between the internal carotid artery and the cavernous sinus.

Table 1 Classification schemes for DAVFs

Venous drainage pattern	Cognard	Borden	Lalwani
Antegrade flow into dural venous sinus	I	I	1
Retrograde flow into dural venous sinus	IIa	I	2
Antegrade flow into dural venous sinus with retrograde flow into cortical vein(s)	IIb	II	3
Retrograde flow into dural venous sinus and cortical vein(s)	IIa + b	II	3
Direct cortical venous drainage without venous ectasia	III	III	3
Direct cortical venous drainage WITH venous ectasia	IV	III	4
Spinal perimedullary venous drainage	V		

Location/Anatomy

The transverse/sigmoid sinuses are the most common sites for DAVFs. Patients typically present with pulse-synchronous tinnitus and headaches, although roughly 10 % will present with acute intracranial hemorrhage (Kirsch et al. 2009; Fig. 1). Spontaneous regression has been reported but is relatively rare (Luciani et al. 2001). The arterial supply to transverse/sigmoid DAVFs is typically from transmastoid branches of the occipital artery, branches of the middle meningeal artery, posterior auricular artery, meningeal branches of the ascending pharyngeal artery, posterior meningeal branches of the ipsilateral vertebral artery, and tentorial branches of the meningohypophyseal trunk (Halbach et al. 1987a). Occasionally, the

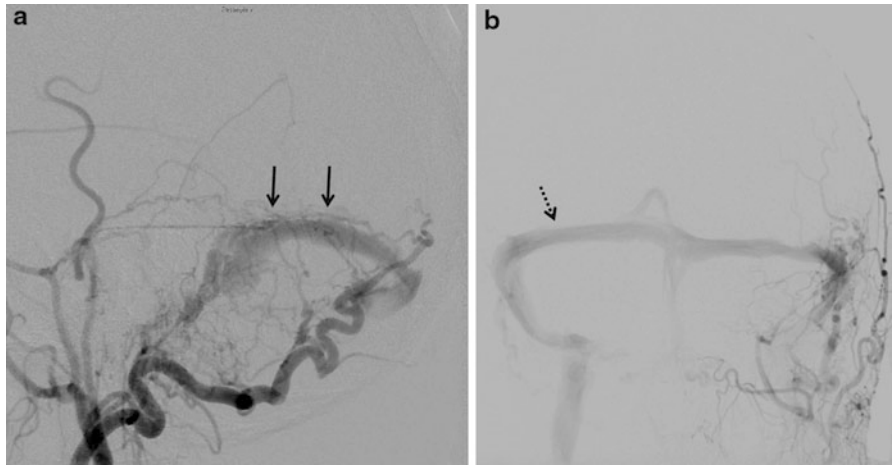

Fig. 1 Seventy-three-year-old male presented with left-sided pulse-synchronous tinnitus. On the suspicion of a DAVF, digital subtraction cerebral angiography was performed. Lateral (**a**, arterial phase) and AP (**b**, arterial phase) views from selective left external carotid arteriography demonstrated hypertrophy and arteriovenous shunt vascularity arising from the occipital, posterior auricular and ascending pharyngeal branches of the left external carotid artery in addition to small branches of the left middle meningeal artery. They converged upon an arteriovenous fistula within the wall of the left transverse sinus (*solid arrows*) and remnant of the left sigmoid sinus (the patient had prior left sigmoid sinus and jugular vein thrombosis). There was retrograde flow in the left transverse sinus crossing midline with drainage to the torcular Herophili and right transverse sinus (*dashed arrow*). No cortical venous drainage was seen. Complete cure was obtained following uncomplicated endovascular coil embolization via the transvenous approach

transverse sinus can be occluded on both ends, resulting in a trapped sinus with exclusive cortical venous drainage. When planning endovascular treatment, particular attention must be paid to direction of flow within the vein of Labbe, as well as its point of insertion into the transverse/sigmoid sinuses. Occlusion at its origin is only safely accomplished if flow is retrograde. If flow is antegrade, occlusion can exacerbate venous hypertension, leading to stroke with aphasia or hemorrhage (Santillan et al. 2013; Fig. 2).

Following the transverse/sigmoid sinus, the cavernous sinus is the next most common site of DAVFs. Common clinical signs and symptoms include chemosis, proptosis, arterialized conjunctival veins, ophthalmoparesis, retro-orbital pain, elevated intraocular pressure, and diminished visual acuity. Cavernous sinus DAVFs frequently receive blood supply from the meningohypophyseal trunk, the inferolateral trunk, McConnell's capsular arteries, the middle and accessory meningeal arteries, the ascending pharyngeal artery, and the arteries of the foramen rotundum and pterygoid canal (Meyers et al. 2002). Supply from the contralateral carotid artery may also occur. Venous drainage is highly variable and depends on several factors including the preexisting venous anatomy, fistula flow rate, and duration of disease (Stiebel-Kalish et al. 2002; Suh et al. 2005).

Superior sagittal sinus DAVFs are relatively rare representing approximately 10 % of cranial fistulae (Fig. 3). They typically present with intracranial hemorrhage or progressive neurological deficits; bleeding rates of up to 40 % have been reported (Kurl et al. 1996; Bertalanffy et al. 2001). Lesions in this location usually involve the mid to posterior third of the superior sagittal sinus and draw their arterial supply from branches of the bilateral middle meningeal arteries. Small contributions from the falcine branches of the ophthalmic arteries and the posterior meningeal arteries can also be seen. Because of their unique midline location, multiple arterial feeders, and cortical venous drainage, superior sagittal sinus DAVFs often require aggressive forms of therapy (Halbach et al. 1988).

First reported by DAVFs of the anterior cranial fossa (ethmoidal) represent a distinct subgroup of DAVFs with a high incidence of intracranial

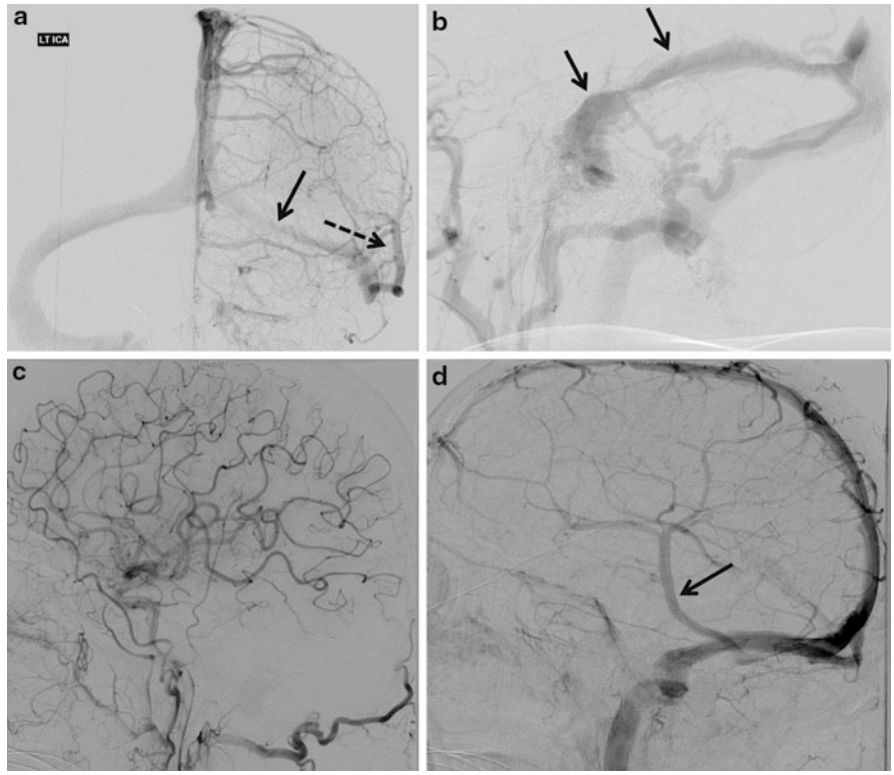

Fig. 2 Sixty-seven-year-old male with left-sided pulse-synchronous tinnitus and severe left-sided headache. Left internal carotid arteriography (**a**, venous phase) demonstrated retrograde flow in the left transverse sinus (*solid arrow*) crossing midline to the right. The left vein of Labbe (*dashed arrow*) drained in antegrade fashion to the left transverse sinus. Left external carotid arteriography (**b**, arterial phase) showed hypertrophy of multiple external carotid branches with an arteriovenous shunt converging on a remnant of the left sigmoid sinus (*solid arrows*). Left common carotid arteriography following transvenous coil embolization (**c**, arterial phase; **d**, venous phase) showed complete occlusion with arrest of all arteriovenous shunt. The left vein of Labbe (*solid arrow*) and transverse sinus remained patent. When planning endovascular treatment of sigmoid/transverse sinus DAVFs, particular attention must be paid to direction of flow within the vein of Labbe, as well as its point of insertion into the transverse/sigmoid sinuses. Occlusion at its origin is only safely accomplished if flow is retrograde. If flow is antegrade, occlusion can exacerbate venous hypertension, leading to stroke with aphasia or hemorrhage

hemorrhage (Fig. 4). Relatively rare, they constitute less than 10 % of all DAVFs (Halbach et al. 1990; Martin et al. 1990; Gliemroth et al. 1999; Lv et al. 2008). They are more common in men, in contrast to the distribution in other locations of DAVFs. Anterior cranial fossa DAVFs almost universally involve the dura in the region of the cribriform plate and the anterior falx. The principal arterial supply comes from the ipsilateral anterior and posterior ethmoidal arteries of the ophthalmic arteries. Additional supply through anastomoses with the contralateral anterior ethmoidal artery as well as ethmoidal branches of the internal maxillary artery, middle meningeal artery, and superficial temporal artery may also exist (Martin et al. 1990; Gliemroth et al. 1999; Lv et al. 2008). Drainage is primarily into pial veins of the anterior frontal lobe which ultimately drain into the superior sagittal sinus or cavernous sinus. Large venous aneurysms (or varices) are an extremely common finding and appear to be the source of hemorrhage (Martin et al. 1990; Gliemroth et al. 1999). Hemorrhage, with reported rates as high as 91 %, may be massive and life threatening. As a result, anterior cranial fossa DAVFs should be treated even if asymptomatic at the time of diagnosis (Reul et al. 1993; Lawton et al. 1999).

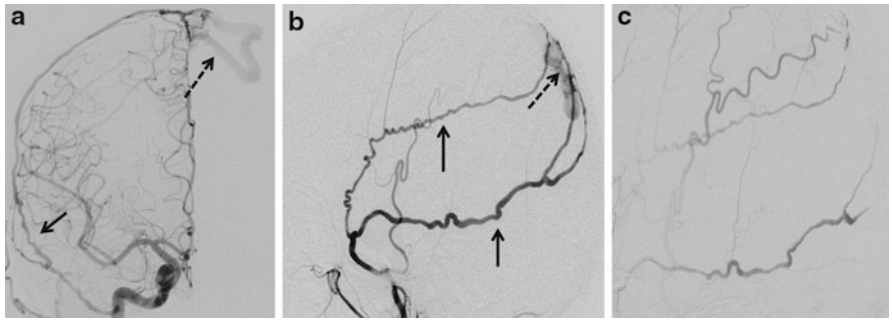

Fig. 3 Sixty-two-year-old male with sudden onset of severe headache. Noncontrast head CT demonstrated extensive posterior fossa subarachnoid hemorrhage. On the suspicion of a cerebral aneurysm, digital subtraction angiography was performed. Right common carotid arteriography (**a**, arterial phase) demonstrated hypertrophy of the right middle meningeal artery (*solid arrow*) with arteriovenous shunting occurring at the level of the superior sagittal sinus, with drainage to a left parasagittal cortical vein (*dashed arrow*). Selective left external carotid arteriography (**b**, lateral, arterial phase) showed rapid arteriovenous shunting arising from anterior and posterior division branches of the left middle meningeal artery (*solid arrows*) converging on a left parasagittal cortical vein fistula. Venous drainage proceeded via dilated and aneurysmal cortical vein (*dashed arrow*) to the adjacent sagittal sinus. The patient subsequently underwent embolization and occlusion of left parasagittal cortical vein fistula using Onyx (eV3 Neurovascular Inc., Irvine, CA). Left external carotid arteriography post embolization (**c**, arterial phase) showed stagnating antegrade flow in the left middle meningeal branch vessels. This treatment proved to be a durable cure

Tentorial dural arteriovenous fistulas represent another unique subset of intracranial DAVFs. These fistulas occur in the dura of the tentorium and its attachments rather than in the sinus wall. Like ethmoidal DAVFs, tentorial DAVFs are relatively rare, accounting for 4–8 % of intracranial fistulae. They are associated with an aggressive natural history, often presenting with hemorrhage or progressive neurological deficits (Zhou et al. 2007; Khan et al. 2009). Several reports have demonstrated tentorial DAVFs manifesting with rapidly progressive myelopathy because of spinal perimedullary venous drainage (Khan et al. 2009; Wrobel et al. 1988). Arterial supply is usually multiple with contributions from the occipital artery, posterior meningeal artery, marginal tentorial artery, middle meningeal artery, meningeal branches of pial arteries, and branches of the external carotid artery. Bilateral internal and external carotid and vertebral angiography should be performed in all patients with tentorial DAVFs to elucidate the potentially complex arterial supply (Khan et al. 2009). Venous drainage is usually through the superior petrosal sinus or through the pontine, perimesencephalic, and basal veins. Most tentorial DAVFs are Borden type II or III lesions because of the presence of retrograde leptomeningeal venous drainage (Grisoli et al. 1984; Tomak et al. 2003; Byrne and Garcia 2013).

Diagnostic Imaging

Although cross-sectional imaging modalities are neither sensitive nor specific for the detection and risk stratification of DAVFs, noninvasive imaging should be the initial study of choice, and pertinent findings must be recognized. Noncontrast head CT can demonstrate areas of hypodensity representing edema or venous ischemia, as well as acute/subacute intracranial hemorrhage. Enlarged pial veins can sometimes be identified as hyperdense tubular structures. Venous varices and sinus thrombosis may also be detected. Proptosis, increased attenuation of intraorbital fat, extraocular muscle enlargement, superior ophthalmic vein enlargement, and enlargement of the cavernous sinus can be seen on noncontrast CT in patients with cavernous sinus DAVFs. Contrast-enhanced head CT can show enhancement of refluxing cortical veins. CT angiography has been shown to demonstrate the feeding artery,

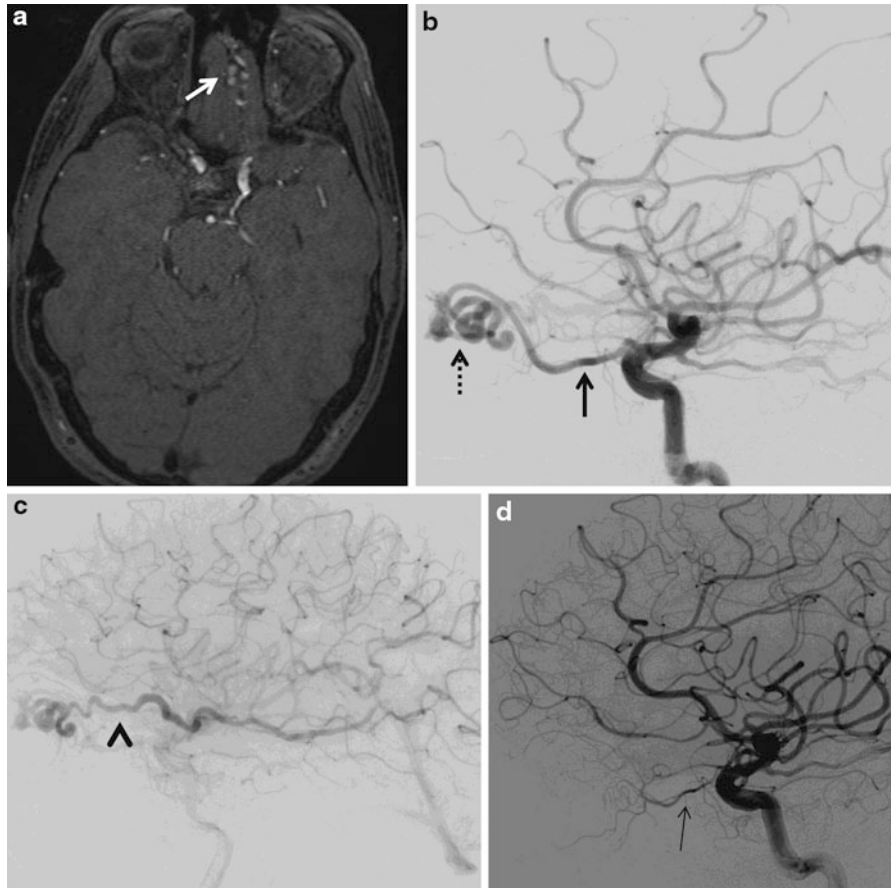

Fig. 4 Forty-one-year-old male presented with severe left-sided frontal headache. MR angiography (**a**) revealed multiple enlarged ethmoidal arteries on the left (*solid arrow*). On suspicion of an ethmoidal dural fistula, catheter angiography was performed. Left internal carotid arteriography (**b**, arterial phase; **c**, late arterial phase) demonstrated hypertrophy of the ophthalmic artery with rapid arteriovenous shunting to dilated veins (*dashed arrows*) and early retrograde filling of the superior ophthalmic vein (*arrowhead*). The patient subsequently underwent transarterial embolization using Onyx. Left internal carotid arteriography (**d**, arterial phase) post embolization demonstrated complete occlusion of the fistula. The left ophthalmic artery (*solid arrow*) had decreased in caliber following occlusion of the fistula

the pattern of venous drainage, and aid localization in all types and grades of DAVFs (Lee et al. 2010; Koc et al. 2013; Fig. 5). It is also helpful in treatment planning by defining the DAVF relative to surrounding brain and skull anatomy. Benign DAVFs, those without cortical venous reflux, are nearly always occult on noncontrast head CT.

On MR imaging, enlarged pial vessels, occluded dural sinuses, and cerebral and cerebellar white matter edema may be found in the presence of cortical venous reflux and venous hypertension (Willinsky et al. 1994; Fig. 6). Enhancement along the peripheral margins of white matter T2 hyperintensity has also been reported (Lee et al. 2003). With time-resolved 3D contrast-enhanced MR angiography, early venous filling of a sinus or cortical vein was found to be reliable signs in detecting DAVFs, in both diagnostic and follow-up imaging (Meckel et al. 2007). However, this same technique is less effective at detecting mild venous reflux and delineation of small feeding arteries compared with digital subtraction angiography (DSA). Additionally, with a reported

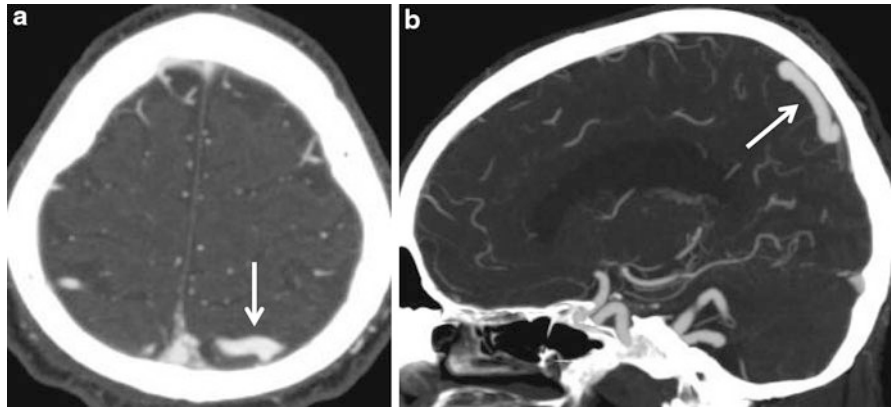

Fig. 5 Sixty-two-year-old male presented with extensive posterior fossa subarachnoid hemorrhage. CT angiography was performed to elucidate the source of bleeding. Select axial (**a**) and sagittal (**b**) images from that study showed a large, enhancing tubular structure over the medial left parietal convexity (*solid arrows*), concerning for an enlarged cortical vein. Digital subtraction angiography was performed confirming the presence of a left parasagittal cortical vein arteriovenous fistula. It was subsequently embolized using Onyx (eV3 Neurovascular Inc., Irvine, CA)

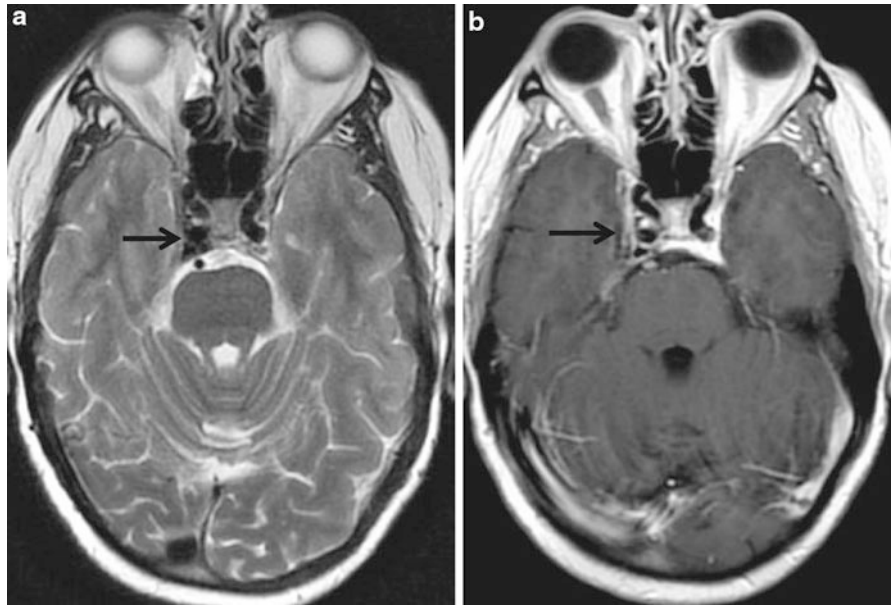

Fig. 6 Sixty-two-year-old woman with headache and diplopia for 9 months. MRI brain with contrast was performed to evaluate. Multiple prominent serpiginous signal flow voids were noted in the right cavernous sinus on the axial T2W (**a**) and T1W (**b**) post contrast images (*solid arrows*). Digital subtraction angiography was subsequently performed demonstrating a type D2 arteriovenous fistula of the right cavernous sinus. Uncomplicated transvenous coil embolization and occlusion were performed

detection rate of only 50 %, and a poor negative predictive value, MR angiography cannot substitute for high-quality DSA (Cohen et al. 2009; Koenigsberg 1996).

DSA remains the gold standard for the diagnosis, risk stratification, and treatment planning for DAVFs. Evaluation should include selective injections of the internal carotid arteries, external

carotid arteries, and vertebral arteries. When the suspected lesion is at the craniocervical junction, selective injections of the thyrocervical and costocervical arteries should be performed. Superselective injections are often required to adequately observe the arterial supply and delineate any anastomoses between the arterial feeders and arteries of the brain, orbit, or cranial nerves. High frame rate, magnified, and oblique views should be obtained to document the exact anatomy of each fistula. Meticulous visualization of the venous system is critical for planning treatment. The presence or absence of cortical venous reflux must be documented. Subtle findings, including pial or medullary collateral veins, focal regions of delayed circulation, and venous rerouting to the orbit or to transosseous veins, must be sought (Willinsky et al. 1999). Tortuous, dilated veins seen over the surface of the brain indicate long-standing venous hypertension. This so called "pseudophlebitic" pattern tends to be associated with an aggressive presentation and retrograde leptomeningeal venous drainage (Willinsky et al. 1999).

Clinical Presentation

The clinical presentation of DAVFs is highly variable, depending on intracranial location, duration of disease, and pattern of venous drainage (Cognard et al. 1995; Hurst et al. 1998; Lucas Cde and Zabramski 2007). The signs and symptoms can change as the disease progresses. DAVFs have been reported to occur at any age, although the majority present in the fifth and sixth decades of life. They are slightly more common in women. They tend to present with more severe symptoms in men, who have a greater number of DAVFs in locations with cortical venous drainage. Patient age, cause of DAVF, and arterial characteristics do not correlate with symptomatology (Cognard et al. 1995). When a patient presents with an unusual constellation of neurological signs and symptoms, DAVF should be included in the differential diagnosis even in the face of normal noninvasive imaging. Additionally, DAVFs should be considered in patients without an obvious cause of intracerebral hemorrhage or who present with signs and symptoms of venous hypertension.

Signs and symptoms of DAVFs can be divided into those that are "benign," and those that are "aggressive." Benign clinical features include pulse-synchronous tinnitus and orbital symptoms including ophthalmoplegia, proptosis, chemosis, and retro-orbital pain. Benign signs and symptoms are typically seen in DAVFs that lack cortical venous drainage. Thus, Borden type I and Cognard types I and IIa DAVFs are considered benign lesions that almost never present with grave pathology (Awad et al. 1990; Fermand et al. 1987; Davies et al. 1997). The most common symptom, pulse-synchronous tinnitus results from turbulent blood flow through a venous sinus near the auditory apparatus in the petrous bone. It may be possible for the physician to appreciate a bruit in the retroauricular or periorbital region on physical exam. Cavernous sinus fistulas characteristically present with proptosis, chemosis, and ophthalmoparesis (Gandhi et al. 2012; Meyers et al. 2002; Preechawat et al. 2008). Findings that warrant emergency treatment include uncontrollable open-angle glaucoma, subretinal effusion, and retinal detachment. Close interaction with the ophthalmologist is critical in patients with cavernous sinus DAVFs to decide when treatment is warranted.

Intracranial hemorrhage and nonhemorrhagic neurologic deficits are considered aggressive clinical features and are almost always seen in DAVFs with cortical venous drainage. Aggressive DAVFs include Borden types II/III and Cognard types IIb/IIIa + b/III/IV/V lesions. Hemorrhage, which results from the venous outflow, may be subdural, subarachnoid, or intraparenchymal as cortical veins traverse each of these different compartments (Kohama et al. 2010; Deshmukh et al. 2005; Liu et al. 2009). As previously stated, anterior cranial fossa and tentorial DAVFs are more prone to present with hemorrhage than DAVFs in other locations as the former are almost always drained by cortical veins. Nonhemorrhagic neurologic deficits come in two varieties, focal and non-focal. Non-focal signs and symptoms appear to be related to passive venous hypertension and/or venous congestion.

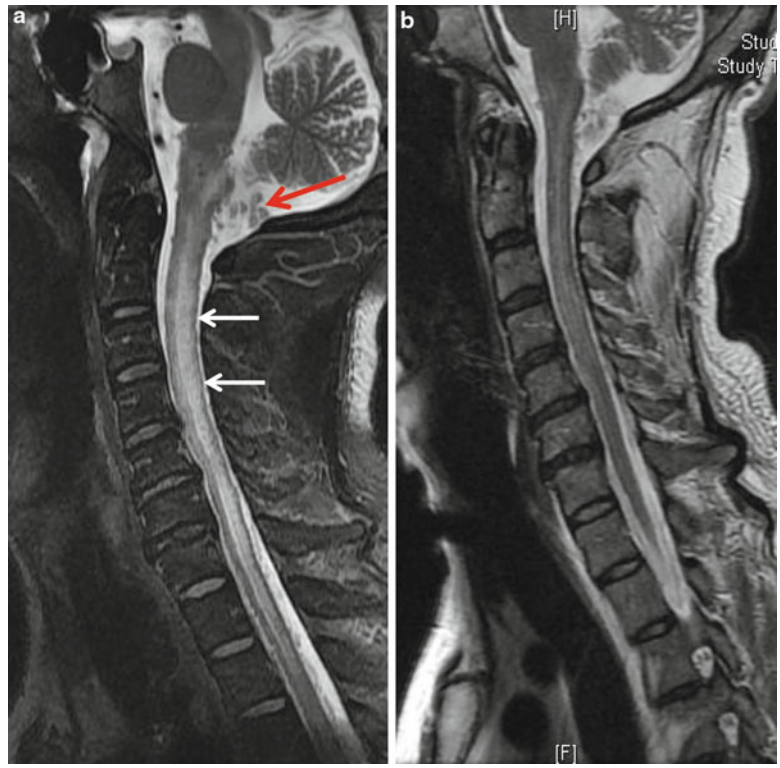

Fig. 7 Forty-one-year-old male presented with rapidly progressive quadriparesis and autonomic failure. Sagittal T2W MR of the cervical spine (**a**) demonstrated serpiginous vessels producing mass along the dorsal aspect of the cervicomedullary junction (*red arrow*) as well as abnormal increased T2 signal and expansion of the cervical spinal cord (*white arrows*). The patient subsequently underwent digital subtraction angiography that revealed a posterior meningeal artery DAVF. It was successfully embolized (see Fig. 8). Cervical spine MRI (**b**) one month after embolization revealed marked improvement in the degree of abnormal spinal cord T2 signal

Focal seizures, transient ischemic attacks, motor weakness, and brainstem and cerebellar symptoms are dependent on the territory of the draining vein or veins (Lasjaunias et al. 1986).

Intracranial DAVFs that drain into perimedullary veins of the spinal cord can cause myelopathy as a result of increased pressure within intramedullary veins leading to reduction in perfusion and microcirculation (Perkash et al. 2002; Kalamangalam et al. 2002; Wiesmann et al. 2000; Akkoc et al. 2006; Renner et al. 2006). Cerebral angiography should be considered in any patient presenting with myelopathy of unknown etiology (Figs. 7, 8). In pediatric patients with high-flow fistulas, cardiac failure and respiratory distress can be seen.

Treatment

The decision to treat a patient with a DAVF requires a multidisciplinary approach involving discussions among interventional neuroradiologists, neurosurgeons, and neurologists and depends on the type of lesion, including location and angiographic risk strata, assessment of clinical presentation, and patient status (age, comorbidities). The risk of treatment should always be weighed against what is known about the natural history of a particular lesion. In general, low-grade fistulae, including Borden I and Cognard I and IIa DAVFs, can be treated conservatively. These benign lesions have an excellent natural history and can typically be followed with MR angiography (Luciani et al. 2001). However, any change in clinical signs or symptoms (even resolution) requires urgent repeat catheter angiography to rule out the development of cortical venous reflux. High-grade lesions, with cortical venous reflux or venous varices, require early treatment to avoid potentially devastating complications of hemorrhage and nonhemorrhagic neurologic deficits. Patients with decreasing visual acuity, visual loss, ophthalmoplegia, intracranial hemorrhage, or infarction require emergent treatment. Re-hemorrhage risks have been reported as high as 35 % within 2 weeks of initial hemorrhage

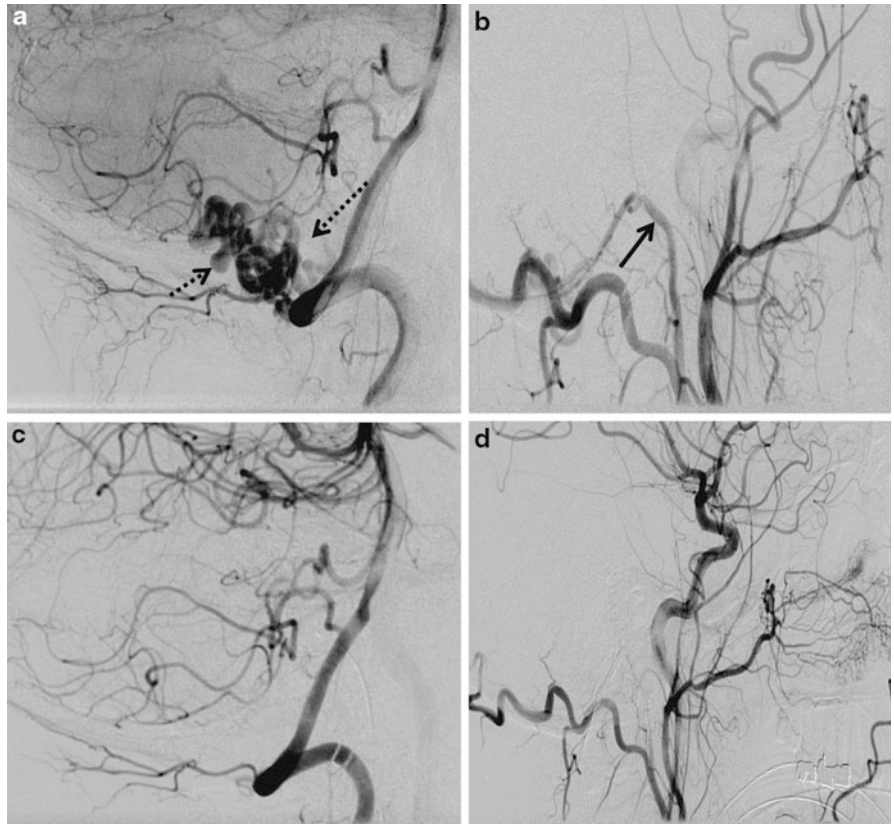

Fig. 8 Forty-one-year-old male presented with rapidly progressive quadriparesis and autonomic failure. Cervical spine MRI revealed serpiginous vessels at the dorsal aspect of the cervicomedullary junction and edema throughout the cervical spinal cord (see Fig. 7). Left vertebral arteriography (**a**, arterial phase) demonstrated mild hypertrophy of the posterior meningeal arteries with rapid arteriovenous shunting to dilated perimedullary veins at the craniocervical junction (*dashed arrows*). Left external carotid arteriography (**b**, arterial phase) showed hypertrophy of the posterior auricular branch of the left occipital artery (*arrow*). Rapid arteriovenous shunting proceeded to the left posterior meningeal artery with early opacification of the dilated perimedullary veins previously seen at the craniocervical junction. Transarterial embolization was performed using Onyx. Left vertebral arteriography (**c**, arterial phase) and left common carotid arteriography (**d**, arterial phase) post embolization demonstrated complete occlusion of the fistula

(Duffau et al. 1999). Low-grade fistulae with debilitating symptoms such as intolerable tinnitus or severe orbital symptoms are candidates for early treatment.

When treatment of a DAVF is indicated, first-line management is endovascular embolization. Microsurgery may be used either in combination with endovascular techniques or when endovascular therapy fails. Although surgical resection or skeletonization of the involved segment of sinus is effective, bleeding complications are not uncommon given the highly vascular nature of DAVFs (Sundt and Piepgras 1983). Stereotactic radiosurgery has been used to treat patients with DAVFs with variable success (Hirai et al. 1998). Improved success rates for radiosurgery have been reported when used as an adjunct to transarterial embolization (TAE). The role of stereotactic radiosurgery remains controversial; whether it should be a first choice of treatment or only a last resort as an alternative to endovascular treatment or surgical resection remains unclear (Hanakita et al. 2012). Compression therapy has been used to treat DAVFs and may still be useful in select patients with low-risk fistulas.

Compression therapy has been used with variable success to treat cavernous sinus and transverse/sigmoid sinus DAVFs and may still be useful in select patients with low-risk lesions (Higashida et al. 1986; Halbach et al. 1987b). This technique requires a compliant, highly motivated patient free of cervical carotid atherosclerotic disease. For patients with cavernous DAVFs, compression of the ipsilateral carotid-jugular sheath is performed for a period of 10 s several times per hour. Compression must only be performed by the patient while seated or lying down using the contralateral hand. The compressing arm should be free to fall if hemispheric ischemia where to occur. If tolerated, the length of compression is gradually increased to 30 s periods several times per hour. With effective compression, there should be marked improvement or total resolution of the bruit. Using this technique Halbach et al. reported complete cure in seven of 23 patients (30 %). No complications occurred (Halbach et al. 1987b). In another series, Higashida et al. reported a cure rate of 30 % using the same technique. At 1-year follow-up no recurrence either clinically or at angiography was reported (Higashida et al. 1986). For transverse/sigmoid sinus fistulae, compression of the occipital artery over the mastoid bone may be effective. Occipital artery compression therapy resulted in a complete cure in two of nine patients (22 %) and improvement in three of nine (33 %) in one series (Halbach et al. 1987b).

Endovascular embolization can be performed via the transarterial route or transvenous route. Complex fistulas may require using a combination of transvenous and transarterial techniques to disconnect the cortical venous reflux and occlude the fistula. TAE involves superselective distal catheterization of dural branches supplying the DAVF with the microcatheter tip in a wedged position within the feeding artery. The goal is to embolize the origin of the draining vein; thus, the embolic agent must penetrate the fistulous connection and proximal aspect of the draining vein. An injection that is too proximal and does not occlude the vein can result in arterial recruitment and persistence of the fistula. An injection too distal may lead to migration within the draining vein(s) resulting in an increased risk of infarction or hemorrhage. Complete cures are sometimes difficult to achieve with TAE because of the existence of feeding arteries that cannot be catheterized and the recruitment of blood supply from collateral arteries. However, TAE may be the only viable option in certain circumstances where transvenous embolization (TVE) is not possible because of limited sinus venous access to the fistula site due to stenosis or thrombosis (Cognard et al. 2008).

Multiple embolic agents can be used to perform TAE including microcoils, polyvinyl alcohol particles, and liquid embolic agents, namely, n-butyl-2-cyanoacrylate (NBCA) and Onyx (eV3 Neurovascular Inc., Irvine, CA). Although particles can reduce shunt flow, they do not produce a durable cure and should not be used as standalone treatment. Particles can be used as a supplement to incompletely treated lesions following glue embolization to reduce the flow in collateral vessels and help to induce thrombosis, or in combination with surgery or radiosurgery (Cognard et al. 2008; Agid et al. 2009). Coils can be used as an adjunct to liquid embolic agents but similar to particles do not typically produce a durable cure when used alone.

The most commonly used embolic agents are the liquid embolics, NBCA and Onyx. NBCA has been extensively used with very good success over the last several decades (Kiyosue et al. 2004). Durable fistula obliteration can be achieved, particularly when the embolic material reaches the fistulous connection. The use of NBCA is predicated on its liquid character, which allows it to penetrate into the venous outflow tract where it ultimately polymerizes into an adhesive, solid material upon contact with blood inducing thrombosis and vessel occlusion. The major disadvantage of NBCA is the somewhat unpredictable nature of its polymerization, often leading to inadequate penetration into the fistula. Another potential disadvantage is the fact that NBCA is an adhesive agent and risks permanent retention of the microcatheter. Glue generally requires a highly experienced operator with the ability to determine the appropriate glue dilution.

Onyx is a relatively new (approved for treatment of cerebral arteriovenous malformations in 2005),

nonadhesive liquid embolic agent that has gained popularity in the treatment of DAVFs. It consists of an ethylene-vinyl alcohol copolymer preparation dissolved in dimethyl sulfoxide (DMSO). Tantalum power is added for radiopaque visualization. Unlike NBCA which polymerizes almost immediately, Onyx has a very slow solidification rate which allows for prolonged, controlled injections. The main advantage of Onyx over NBCA is the ability to administer a large volume of embolic agent via a long-duration injection through 1 pedicle (Panagiotopoulos et al. 2009).

Recent preliminary reports show high curative rates associated with transarterial embolization with Onyx. Nogueira et al. treated 12 consecutive patients with intracranial DAVFs between March 2006 and February 2007 using Onyx. All procedures were performed via a transarterial approach. Complete angiographic cure was seen in 11 of the 12 patients (91.7 %). There was no significant morbidity or mortality, and no patient required follow-up surgery or radiosurgery (Nogueira et al. 2008). Cognard et al. reported on 30 patients who underwent endovascular treatment for DAVFs with cortical venous reflux. Onyx embolization, via a transarterial approach, resulted in complete angiographic cure in 24 cases (Cognard et al. 2008). A retrospective review of the Barrow Neurological Institute endovascular database highlighted 50 patients with 63 cranial DAVFs that were treated with transarterial Onyx embolization. When Onyx was used as the sole embolic agent, 32 of 37 DAVFs (87 %) achieved angiographic cure (Hu et al. 2011). Jiang et al. retrospectively studied five patients with transverse/sigmoid sinus DAVFs in whom transverse sinus packing was attempted with a transarterial approach using Onyx. Embolizations were all performed via the middle meningeal artery and angiographic cure was obtained in all five cases. No complications were seen (Jiang et al. 2011). Stiefel et al. reported a 72 % complete occlusion rate using Onyx, with four complications (Stiefel et al. 2009).

TVE pioneered by Mullan has been shown to have a high likelihood of cure and is often used to treat transverse/sigmoid sinus and cavernous sinus DAVFs (Stiefel et al. 2009; Roy and Raymond 1997). It is usually performed via the transfemoral approach, although jugular vein puncture, facial vein puncture, superior ophthalmic vein puncture, or even transcranial approaches may be necessary (Naito et al. 2002; Houdart et al. 2002; Jiang et al. 2013). On occasion, a multidisciplinary surgical and endovascular approach must be utilized to cure deep lesions with limited surgical or transvascular access (Zink et al. 2004; Kong et al. 2007). TVE is performed by retrograde catheterization of the involved dural sinus or cortical vein followed by deposition of coils and/or liquid embolic material adjacent to the arteriovenous shunt. The preferred technique at our institution is to initially deploy a detachable coil(s) to form a scaffolding or anchoring basket to prevent migration of subsequent coils. Detachable coils allow for excellent control of coil deposition such that a coil can be placed at the precise location within the sinus. Pushable fibered coils, which are more thrombogenic and much less expensive, are then added to achieve complete occlusion of the venous segment (Fig. 9).

The goal of treatment is occlusion of the fistula and/or disconnection of leptomeningeal or cortical reflux while preserving normal venous drainage. Careful examination of the venous drainage pattern before embolization is crucial to determine if the patient will tolerate sinus occlusion. TVE is safest when the diseased sinus segment has minimal contributions to normal venous outflow and can be completely occluded (Gandhi et al. 2012). The presence of a cortical vein that uses the involved sinus for drainage should suggest a high risk of venous infarction or worsening of neurological deficit if the sinus is occluded. In these circumstances precise identification of the fistula is essential to avoid potential complications (Davies et al. 1997; Gandhi et al. 2012). Additionally, occlusion of major cortical veins such as Labbe, Trolard, internal cerebral veins and the vein of Galen should also be avoided, unless it is clear that they do not serve to drain parenchymal blood. Partial dural sinus embolization should be avoided as shunt flow into the normal cerebral venous pathways can worsen cortical venous drainage (Davies et al. 1997). The ability to

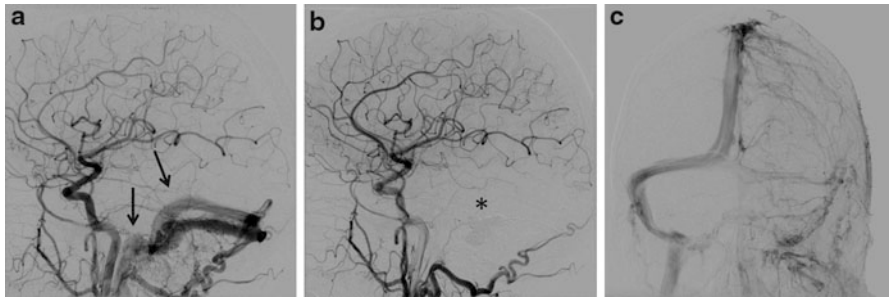

Fig. 9 Seventy-three-year-old male patient presented with left-sided pulse-synchronous tinnitus and headache. Left common carotid arteriography (**a**, arterial phase) showed arteriovenous shunting arising from multiple branches of the left external carotid artery converging on the remnant of the left sigmoid sinus (left sigmoid sinus and jugular veins previously thrombosed). There was retrograde flow crossing midline via the left transverse sinus to the patent right transverse sinus and jugular vein (*solid arrows*). The patient subsequently underwent uncomplicated endovascular occlusion and eradication of the arteriovenous fistula via the transvenous approach using multiple platinum and fiber platinum detachable and pushable coils. Left common carotid arteriography (**b**, arterial phase; **c**, venous phase) following embolization showed no arteriovenous shunt vascularity (*coil mass). Major dural sinuses and cortical venous structures of the posterior fossa remained patent with the exception of the site of endovascular occlusion

close a fistula in 1 session as well as the relative ease with which one can gain retrograde venous access makes TVE embolization very appealing. This technique is particularly useful for DAVFs with multiple arterial feeders that are either too small or too tortuous to treat with TAE. Risks of TVE include vessel perforation, cerebral infarction, intracranial hemorrhage, and focal neurological deficits related to changes in the venous drainage pattern (Gandhi et al. 2012).

Conclusion

Intracranial DAVFs are rare pathologic lesions accounting for 10–15 % of intracranial arteriovenous shunts. Signs and symptoms are highly variable and depend upon several factors including fistula location, duration of disease, and most importantly venous drainage pattern. Symptoms range from pulse-synchronous tinnitus and exophthalmos to cranial nerve deficits, cognitive impairment, venous infarction, intracranial hemorrhage, and even death. DAVFs are dynamic lesions that may either spontaneously regress or progress, and close clinical and radiologic monitoring is mandatory. Treatment should be pursued for all lesions with cortical venous drainage or intolerable symptoms. Management decisions require a multidisciplinary approach involving discussions among interventional neuroradiologists, neurosurgeons, and neurologists.

Cross-References

- ▶ Embolization of Cerebral Arteriovenous Malformations
- ▶ General Angiographic Technique in Neuro-Interventional Therapy
- ▶ Spinal Vascular Malformations

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Further Reading

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