Transverse-Sigmoid Sinus Dural Arteriovenous Malformations

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Learning Objectives: *After reading this article, the participant should:*

1. *Have an in-depth knowledge of the etiologic factors and clinical symptoms of transverse-sigmoid sinus dural arteriovenous malformations.*
2. *Have an in-depth knowledge of the radiologic features of transverse-sigmoid sinus dural arteriovenous malformations.*
3. *Have in-depth knowledge of indications for treatment and treatment options of transverse-sigmoid sinus dural arteriovenous malformations.*

Dural arteriovenous malformations (AVM), also commonly described in the literature as dural arteriovenous fistulae, consist of a lesion, or nidus, containing arteriovenous shunting and confined within the dural leaflets. The essential radiologic defining features are the nidus of the malformation and the early appearance of venous structures during the arterial phase of cerebral angiography.

The intracranial arteriovenous malformations are classified on the basis of their arterial supply into pure pial, mixed pial and dural, and pure dural AVMs. Dural AVMs account for 10% to 15% of all arteriovenous intracranial malformations; they account for 6% of supratentorial and 35% of infratentorial arteriovenous malformations. Dural AVMs most commonly involve the transverse sinus (63% of cases), with the nidus invariably located at the transverse-sigmoid sinus junction. There is a female preponderance (61% to 66%) and a slight left-sided preference (60%). Patients usually are in the fifth or sixth decade of life.

Pathogenesis

Dural AVMs are a nosologically heterogeneous group of lesions linked by similar architecture. Theories vary regarding the etiology and pathogenesis of these lesions. Some researchers believe that congenital factors, namely the persistence and enlargement of primitive dural arteriovenous communications that normally involute during fetal development, are causal. Dural AVMs rarely are seen during childhood, however. When they do occur in children, these lesions tend to be complex and bilateral. They are more common in boys, and usually are seen with cardiac failure at presentation and have a high mortality rate (38%).

Most dural AVMs occur in middle-aged patients. However, this fact does not exclude a certain degree of embryological contribution to their pathogenesis. For example, in some patients "spontaneous" transverse-sigmoid sinus dural AVMs have developed without evidence of compromise to flow in the sinus. Nonetheless, acquired factors are believed to play the major role in the etiology of transverse-sigmoid sinus dural AVMs.

Few reports have suggested that sinus thrombosis is secondary or subsequent to occurrence of transverse-sigmoid sinus dural AVMs. However, most of the evidence suggests that thrombosis of the dural venous sinus precedes their appearance. A rate as high as 72% has been reported for sinus thrombosis concomitant with transverse-sigmoid sinus dural AVMs.

A sequence of three stages in the natural history of transverse-sigmoid sinus dural AVMs has been suggested as a possibility: (1) occurrence of sinus thrombosis with engorged

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dural venous collaterals and the opening of embryonic arteriovenous communications; (2) development of arteriovenous shunting, which favors the recruitment of arterial feeders into the nidus, which can lead to secondary venous hypertension; and (3) development of leptomeningeal retrograde venous drainage in the later phase, predisposing to varicose and aneurysmal dilation of the pial veins and a higher possibility of intracranial hemorrhage. The role of venous hypertension in the development of transverse-sigmoid sinus dural AVMs has been experimentally studied in rats and successfully reproduced.

Other factors reported to compromise transverse-sigmoid sinus flow and precede the development of a dural AVM are previous surgery in the area, trauma, sinus thrombosis, infection, tumors, conditions of increased coagulability, rupture of an aneurysm, arterial dysplasia, pregnancy, hormonal factors, and vascular diseases. Transverse-sigmoid sinus dural AVMs appear to be more commonly associated with compromise of the dominant transverse-sigmoid venous sinus.

Clinical Signs and Symptoms

Clinical signs and symptoms are related to the location of the lesion or, more commonly, to the pattern of venous drainage. Pulsatile tinnitus, the most common symptom, often is very disturbing, and may lead to insomnia and significant discomfort and distress. It is one of the most common indications for surgery. More than 40% of all patients presenting with pulsatile tinnitus have an objective bruit. Other symptoms include headache; altered mental status; memory problems; ataxia and neurological deficits related to cranial nerve involvement; hemorrhage (subdural, subarachnoidal, and intracerebral); increased intracranial pressure (marked by headache, nausea, vomiting, papilledema, optic atrophy, and decreased visual acuity); venous outflow obstruction; seizures; vertigo; and hydrocephalus due to the hemorrhage or mechanical CSF obstruction. Table 1 presents the incidence of the different clinical signs and symptoms.

TABLE 1. Clinical Signs and Symptoms Associated With Transverse-Sigmoid Sinus Dural AVMs

Signs and Symptoms	Incidence (%)
Pulsatile tinnitus	67–92
Headaches and intracranial pain	43–50
Increased intracranial pressure	18–36
Seizures	12–20
Hemorrhage (subdural, subarachnoidal, intracerebral)	8–15
Focal neurological signs	5–14
Altered mental status and memory	6–12
Ataxia	7–8
Cranial nerve involvement	7
Vertigo	2

Diagnostic Studies

The primary diagnostic study for transverse-sigmoid sinus dural AVMs is selective cerebral angiography (Figs. 1A, 1B, and 2). The goals are the proper analysis of arterial feeders, localization of the nidus, and verification of the venous drainage pattern. Magnetic resonance imaging (MRI) is helpful in evaluating the cortical venous drainage and associated sinus thrombosis (Fig. 3). Spin-echo and gradient echo MRI images can help differentiate the slow flow in the sinus from sinus thrombosis, or the dilated cortical veins from hemorrhage. Distinguishing between partial and complete occlusion of the transverse-sigmoid sinus may be difficult because of the anterograde flow from the transverse-sigmoid sinus dural AVMs to the sigmoid sinus, and the retrograde flow from the involved transverse sinus toward the torcula Herophili. Magnetic resonance angiography (MRA) and magnetic resonance venography (MRV) are being used with increasing frequency, and can provide most of the information that previously could be obtained only by cerebral angiography.

Arterial Feeders

Cerebral angiography is the best diagnostic study of arterial feeders of transverse-sigmoid dural AVMs. Feeders of transverse-sigmoid sinus dural AVMs come from the external carotid, internal carotid, and vertebral arteries. The most common primary feeder is the occipital artery. Other involved arteries are the posterior auricular artery, the ascending pharyngeal artery, branches of the internal carotid artery (meningohypophyseal trunk, inferolateral trunk), the

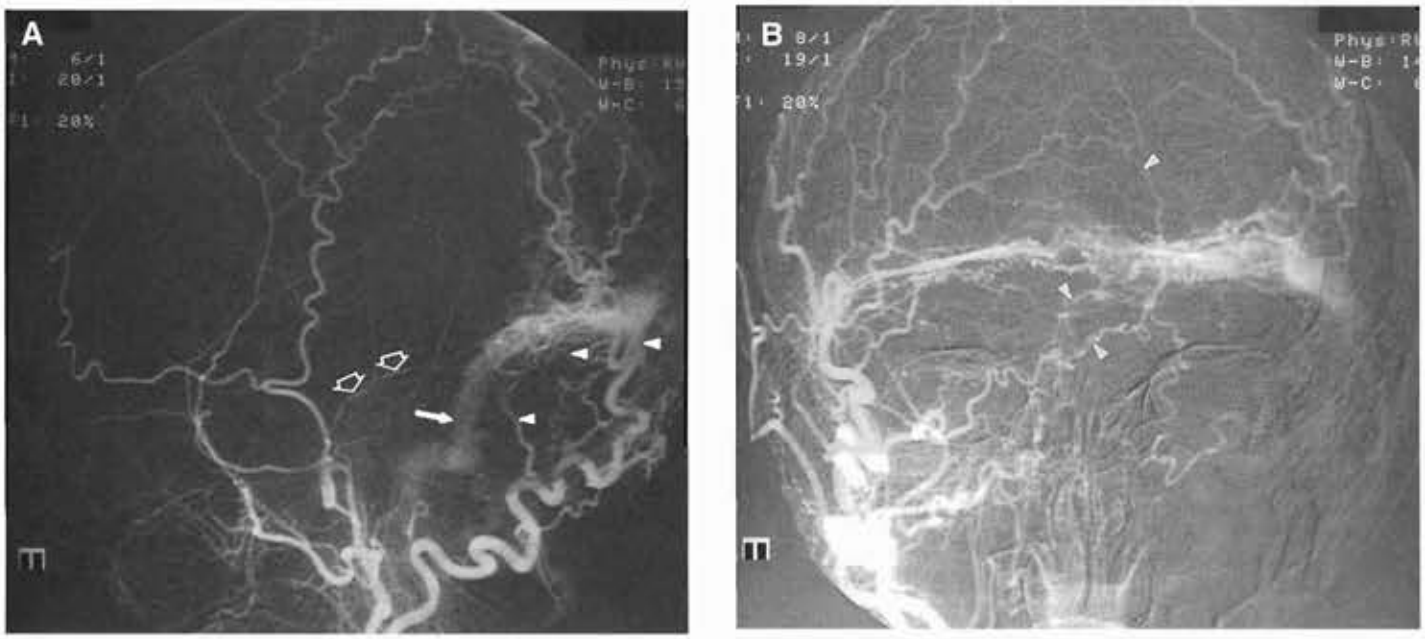

Figure 1. (A) Transverse-sigmoid sinus dural AVM located at the transverse-sigmoid junction and draining into the sigmoid sinus and further into the jugular vein (*arrow*). It is seen in the early arterial phase of external carotid artery injection (lateral view). Feeders to the malformation arise from the branches of the occipital artery (*arrowheads*), dural feeders (*open arrows*), and feeders from the superficial temporal artery. (B) Anteroposterior projection of the early arterial phase of the external carotid artery injection of the same patient. Note the contralateral contribution of the occipital artery branches and the posterior branches of the middle meningeal artery (*arrowheads*).

middle meningeal artery, the superficial temporal artery, and branches of the vertebral artery (the posterior meningeal artery, the muscular branch, and its anastomosis) (Figs. 1A, 1B, and 2). Bilateral feeders are common in more complex dural AVMs (Figs. 1A and 1B), especially mixed pial and dural AVMs. Table 2 lists the vessels that feed transverse-sigmoid sinus dural AVMs.

Venous Drainage

The venous drainage flow pattern of transverse-sigmoid sinus dural AVMs may be normal on angiography (that is, toward the sigmoid sinus and the internal jugular vein), but

Table 2. Arterial Feeders of Transverse-Sigmoid Sinus Dural AVMs

Arterial Feeder (artery)	Frequency (%)
General	
External carotid	89
Internal carotid	51
Vertebral	31
Principal	
Occipital	89–96
Middle meningeal	33–56
Ascending pharyngeal	26
Vertebral artery branches (posterior meningeal, muscular)	21–26
Internal carotid branches (meningohypophyseal, inferolateral trunk)	19–60
Posterior auricular	17–26
Superficial temporal	4

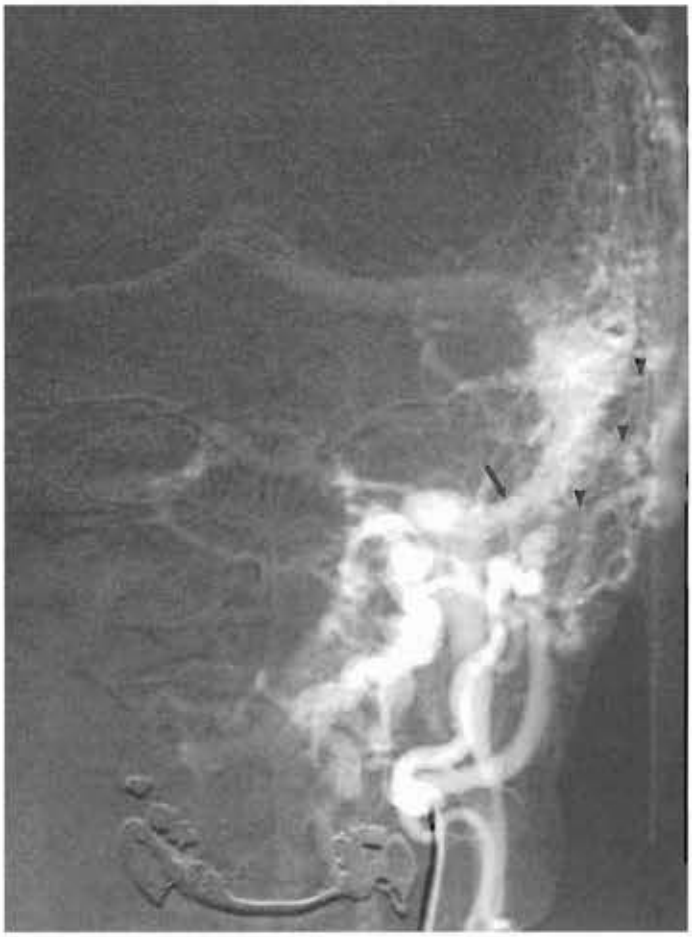

Figure 2. Anteroposterior view of the early arterial phase of external carotid artery injection. Note the branches of occipital artery (*arrowheads*) supplying the transverse-sigmoid sinus dural AVM and the venous drainage into the ipsilateral sigmoid sinus (*arrow*).

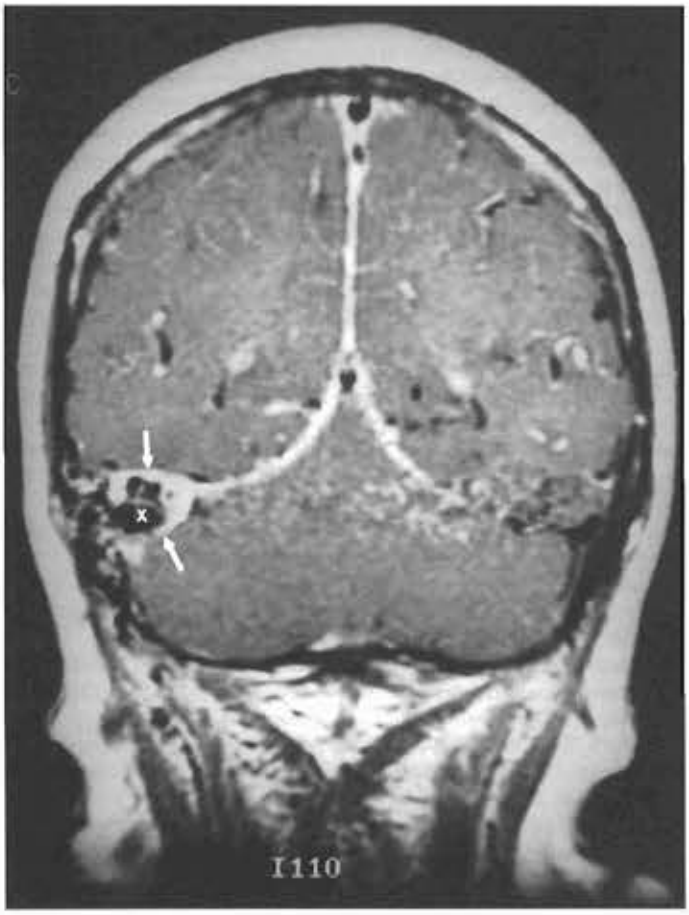

Figure 3. Coronal gadolinium-enhanced MRI of the brain showing a partially thrombosed transverse sinus. The dural AVM enhancement is indicated by the arrows, and the lumen is marked with an X.

more often there is partial compromise of sinus drainage or complete sinus thrombosis. In the latter case, the flow is abnormal, with venous reflux seen in several directions, including the contralateral transverse sinus, the superior and inferior petrosal sinuses, the cavernous sinus, and the vein of Labbe. In such cases, subsequent rise of sinus venous pressure may lead to intracranial hypertension. The increased pressure within the superior sagittal sinus (and its occasional thrombosis) leads to cerebrospinal fluid malabsorption, a hydrostatic type of hydrocephalus that results in a rise in the intracranial pressure and generalized central nervous system dysfunction. The most dangerous phase during the course of a transverse-sigmoid AVM is the development of leptomeningeal retrograde venous drainage, which results in varicose and aneurysmal venous formations. This phase commonly presents with an aggressive clinical course, with increased risk of subdural, subarachnoid, intraventricular, or intracerebral hemorrhage and focal neurologic symptoms. Fortunately, transverse-sigmoid sinus dural AVMs progress to this dangerous phase less often (in only 8% to 10% of cases) than do dural AVMs at other locations.

Treatment

The course of most transverse-sigmoid sinus dural AVMs is benign. Treatment ranges from expectant observation to more involved multimodality treatment interventions. There are reports of spontaneous regression and closure of dural AVMs with presumed thrombosis within the lesion, but this possibility does not necessarily imply a cure. It could be a phase in the dynamic processes that these lesions undergo, because they may later recanalize and recruit new feeders with further enlargement of the fistula. For this reason, spontaneous obliteration of a previously diagnosed transverse-sigmoid sinus dural AVM should not be considered a cure, and the AVM should continue to be observed as a chronic disease process.

When these lesions progress and become more symptomatic, a treatment plan should be initiated. Incapacitating tinnitus is one such reason to initiate a treatment plan. Venous hypertension and evidence of cortical venous reflux are strong indications for initiating a treatment plan even in the absence of significant clinical symptoms, because lesions with such changes have a high potential for causing intracranial hemorrhage.

The goal of treatment should be complete obliteration of the arteriovenous shunting. Endovascular transarterial therapeutic embolization with various thrombogenic materials, including polyvinyl alcohol particles and cyanoacrylate, initially was used as an adjunct to surgery. It embolizes the nidus only incompletely and does not lead to a cure. Later recruitment of nonembolized feeders and enlargement of the fistula are likely. Provocative testing, such as administration of amobarbital, to predict any functional loss from embolizing the vessel before occluding the leptomeningeal feeders may be beneficial. However, transarterial embolization is reserved for very specific situations because of its low potential to lead to a cure. One such situation is that in which the transvenous route is not possible because the patient is considered to be at high surgical risk, and cortical venous drainage and increased intracranial pressure are present. In such a case, transarterial embolization may decrease arterial input across the fistula and also may decrease venous reflux into the pial vessels, thus decreasing the chance of hemorrhage from cortical veins. There are complications related to transarterial embolization, however, including ischemic stroke, which has been reported as a result of embolic material embolizing into the posterior cerebral artery. Venous infarction has been reported to have been caused by embolic material passing retrogradely into the fistula and lodging in the cortical veins.

The transvenous route uses the draining sinus as an access route for obliteration or thrombosis of the nidus of the dural AVM. It leads to a higher rate of cure in most patients when compared with transarterial embolization. There are risks associated with the transvenous approach, however. Pulmonary embolization of the embolic material is one possibility. Incomplete obliteration of the fistula is in itself a potential cause of morbidity due to compromise of the cortical venous drainage, leading to venous hypertension and the increased possibility of hemorrhage. The goal of treatment should not stop short of complete obliteration of the fistula. In most patients this is achieved by complete obliteration of the sinus itself. When the decision to obliterate the sinus is made, it is important to confirm that the

other sinus is patent; if it is not patent, open surgical treatment is a better choice.

The surgical approach currently is used as part of a multimodality treatment plan that includes both transarterial and transvenous embolization. It is used more commonly for complex lesions with multiple arterial feeders. All of the tentorial and dural feeders to the fistula are coagulated and cut, and the transverse sinus is resected. Surgery also is indicated when sacrifice of the sinus could compromise the cortical venous drainage, as in the absence of the contralateral sinus. In this case, resection of the fistula involving the dura of the tentorium and skeletonizing the sinus without sacrificing it becomes an option. Preoperative embolization of the fistula is helpful in reducing blood loss during surgery, especially during the exposure steps of the craniotomy. Use of the high-speed diamond drill can be very helpful in cutting down on blood loss arising from the arterialized venous drainage that has developed through the bone diploë. Even though surgery is curative, in the majority of patients it is most commonly considered when the endovascular option is not possible or is considered very risky.

Stereotactic radiosurgery has recently been used in the treatment of tentorial dural AVMs. However, although it is not frequently considered in the treatment plan of dural AVMs, it can potentially be used for treating transverse dural AVMs in select patients.

Readings

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1. The persistence and enlargement of embryonic dural arteriovenous communications is one of the suggested factors in the etiology of transverse sinus dural AVMs.
True or False?

2. Transverse sinus dural AVMs are the most common type of dural AVM.
True or False?

3. Thrombosis of the dural venous sinus is seen in more than two thirds of patients with transverse sinus dural AVMs.
True or False?

4. Pulsatile tinnitus is one of the most common symptoms in patients with dural AVMs.
True or False?

5. The posterior auricular artery usually is the primary feeder of transverse-sigmoid sinus dural AVMs.
True or False?

6. Generalized intracranial hypertension usually does not occur with transverse-sigmoid sinus dural AVMs.
True or False?

7. A transverse sinus dural AVM with evidence of retrograde leptomeningeal venous drainage is a predictor of a more aggressive clinical course.
True or False?

8. The presence of cortical venous reflux in transverse-sigmoid sinus dural AVMs is not an indication to initiate treatment in the absence of progressive clinical symptoms.
True or False?

9. Endovascular transarterial embolization is the treatment of choice in transverse-sigmoid sinus dural AVMs because of its high rate in achieving cure.
True or False?

10. The direct surgical approach no longer has a role in the treatment of transverse-sigmoid sinus dural AVMs.
True or False?