Dural Arteriovenous Malformations of the Transverse/Sigmoid Sinus Acquired from Dominant Sinus Occlusion by a Tumor: Report of Two Cases

Kenan I. Arnautović, M.D., Ossama Al-Mefty, M.D., Edgardo Angtuaco, M.D., Lori Jo Phares, R.N.
Departments of Neurosurgery (KIA, OA-M, LJP) and Radiology (EA), University of Arkansas for Medical Sciences, Little Rock, Arkansas

OBJECTIVE AND IMPORTANCE: Debate continues regarding the pathogenesis of dural arteriovenous malformations (dAVMs). The prevailing theory is that dAVMs are acquired lesions that occur after thrombosis of the dural venous sinus.

CLINICAL PRESENTATION: We report unique cases of two patients having different tumors (one meningioma and one glomus jugulare paraganglioma) that occluded the ipsilateral transverse and sigmoid sinuses, respectively, and were associated with dAVMs. In each patient, the occluded venous sinus was the dominant sinus.

CONCLUSION: Our experience with these patients supports the hypothesis that dAVMs are acquired and induced lesions that may occur after sinus occlusion. We suggest that the occlusion of the dominant transverse/sigmoid sinus is a major contributing factor to the development of dAVMs because of the inability of the contralateral (nondominant) sinus to handle the venous flow from the obstructed (dominant) side. (Neurosurgery 42:383-388, 1998)

Key words: Dominant dural venous sinus, Dural arteriovenous fistula, Dural arteriovenous malformations, Glomus jugulare paraganglioma, Meningioma, Transverse/sigmoid sinus

Dural arteriovenous malformations (dAVMs) (also commonly described in the literature as dural arteriovenous fistulas) consist of arteriovenous shunts of blood confined within dural leaflets. Essential defining features are the nidus of the malformations and the early appearance of venous structures during the arterial phase of angiography (3). The most common location of dAVMs is in the transverse and sigmoid venous sinuses (3, 20), with the nidus invariably localized at the transverse/sigmoid junction (22).

The cause and pathogenesis of dAVMs remain unclear. Reports of congenital dAVMs have been published (26), and cases of dAVMs without a compromised venous sinus have been reported (4, 31, 32, 34, 41). A few reports have suggested that sinus thrombosis is either a secondary (subsequent to) or a "two-directional process" (1, 2, 12, 31, 33). Nonetheless, the prevailing thought is that the appearance of dAVMs is preceded by compromise of the dural venous sinus (3, 6, 8, 13, 15, 16, 19, 21, 22, 25, 29, 35, 37).

Occlusion of the transverse/sigmoid sinus occurs in a variety of clinical cases. Such occlusion, however, has relatively rare association with dAVMs. We recently encountered two patients with different tumors that occluded the transverse/sigmoid venous sinus and were associated with dAVMs. In these patients, two factors were present that we think were major contributors to the evolution of the dAVMs: 1) tumor occlusion of the dominant draining transverse/sigmoid sinus; and 2) a small contralateral, nondominant venous sinus unable to handle the venous drainage from the occluded side.

CASE REPORTS

Patient 1
Tentorial meningioma

A 63-year-old woman had a several-year history of headache, localized pain in the left orbit, left paranasal sinuses, and constant tinnitus in her left ear, which produced an objective bruit. Preoperative magnetic resonance imaging (MRI) revealed an enhancing extra-axial tumor along the dural leaflet of the left subtentorial region, with occlusion of the transverse sinus (Fig. 1A). Coronal reprojec- tion of a magnetic resonance venogram (MRV) study showed flow signal loss along the left proximal transverse sinus, with good visualization of the distal transverse/sigmoid sinus. Prominent signal changes were noticed along the outer surface of the sigmoid sinus, which represents flow along the dural surface of the sinus. Drainage to the right transverse/sigmoid sinus was poorly visualized. Drainage of the dAVMs was through the left distal transverse and sigmoid sinus (Fig. 1B).

A left external carotid angiogram demonstrated supply to the dAVMs from the right occipital artery (Fig. 1C).

Partial embolization of the feeding arteries of the dAVMs (left and right occipital arteries and the posterior branches of the middle meningeal artery) was performed preoperatively. After a suboccipital craniotomy and a retrosigmoid dural incision, a meningioma was observed adhering to the tentorium along the medial aspect of the transverse sinus. The lesion invaded and occluded the sinus. The entire tumor was...
FIGURE 1. Tentorial meningioma. A, coronal T1-weighted magnetic resonance image showing an enhancing tumor (*) at the left tentorium that extends toward the left transverse/sigmoid sinus (arrow). B, coronal reprojection of a two-dimensional “time-of-flight” MRV showing the occlusion of the left (dominant) transverse sinus (arrow) by the tumor. There is an increased flow in the left distal transverse/sigmoid sinus with multiple rounded flow signals along the dural leaflets of the left distal transverse/sigmoid sinus (arrowheads). Note the poor flow of the contralateral (right) transverse/sigmoid sinus (large arrowhead). C, left (ipsilateral) external carotid arteriogram (lateral projection) showing enlarged branches of the occipital artery (small arrowheads) and the posterior branches of the middle meningeal artery (large arrowheads) supplying the dAVMs (*). D, follow-up T1-weighted postcontrast magnetic resonance image showing radical resection of the tumor. Note also a fat graft (*) in the tumor's bed. E, follow-up coronal reprojection of a two-dimensional time-of-flight MRV showing radical excision of dAVMs without evidence of recurrence (left side).

The dural meningioma was surgically removed, and its base was coagulated. The tentorium had a bluish color, and an abnormal area of blood collection representing the dAVMs was continuous with the tumor along the posterior tentorial margin. The area was coagulated with a bipolar cautery in a stepwise fashion, and the dAVMs were completely excised. In addition, the entire tentorium on the left side was excised, including all residual tumor and the dAVMs.

After the surgery and during the follow-up period (40 mo), the patient was free from headaches and tinnitus. Follow-up postcontrast T1-weighted MRI (Fig. 1D) was performed and an MRV (Fig. 1E) was obtained on two occasions, which confirmed the radical excision of the tumor and dAVMs.

Patient 2
Jugular foramen paraganglioma

A 57-year-old woman had a several-year history of vertigo. She also had a several-month history of dizziness and functional hearing loss and weakness of the tongue on the right side. In addition, she had intermittent numbness on her left side, a right-sided paresis of the lower cranial nerves (Cranial Nerves IX-XII), and a mild contralateral hemiparesis. An MRI study revealed a large, enhancing tumor of the right jugular foramen that extended posteriorly into the cerebellopontine angle and inferiorly along the carotid sheath (Fig. 2A). An MRV disclosed associated dAVMs, with the ipsilateral dominant sigmoid sinus obstructed. There was retrograde flow from the transverse sinus to the small, nondominant, contralateral (left) transverse/sigmoid sinus (Fig. 2B). The tumor was supplied by the right ascending pharyngeal artery, the stylomastoid artery from the right posterior auricular, and the caroticotympanic artery from the right internal carotid artery (ICA). The associated dAVMs were supplied by the mastoid branches of the right occipital artery, the mastoid branches of the right middle meningeal artery, the muscular branches of the vertebral artery, and the tentorial branches of the ICA (Fig. 2, C and D).

The extracranial blood supply to the tumor and the dAVMs was partially...
DISCUSSION

The lesions were approached through the right transtemporal approach. During the first stage of surgery, the extracranial portions of the lower cranial nerves (Cranial Nerves IX–XII) and the vascular structures of the neck were dissected. The ascending pharyngeal, posterior auricular, and occipital arteries were coagulated. The seventh nerve was skeletonized and transposed from its canal. The paraganglioma invaded the wall of the carotid canal in the petrous bone, which was drilled away. It also invaded and occluded the lumen of the sigmoid sinus, which was ligated distal to the tumor invasion and proximal to the mastoid emissary vein. The extradural portion of the tumor was then resected. With continued exposure of the extradural tumor, a significant amount of bleeding was encountered along the exposed dura from the associated dAVMs. The dAVMs were carefully identified, the arterial feeders were localized and meticulously coagulated, and the dAVMs were excised.

During the second stage of surgery, the intradural portion of the tumor was observed to grossly involve the IXth and Xth cranial nerves. The tumor and the dAVMs were removed, the dura involved by the tumor or dAVMs was excised, and duraplasty was performed.

Immediately after surgery, the patient had palsy of the lower cranial nerves, and a gastrostomy tube was placed. Vocal cord medialization was also performed. A gold weight was placed on the patient’s right upper eyelid to overcome seventh nerve palsy. The patient gradually resumed oral feedings, her mild hemiparesis resolved, and she was able to walk. At the most recent follow-up examination (28 mo after surgery), she had House Grade III seventh nerve function on the right side, mild hoarseness, and functional hearing loss on the right side. Other preoperative neurological symptoms were resolved. Follow-up postcontrast T1-weighted MRI confirmed radical tumor excision (Fig. 1E).

Causes and Pathogenesis

The statement that dAVMs are a nosologically heterogeneous group of...
lesions linked by their architecture (19) highlights the diversity of opinion about the cause and pathogenesis of these lesions. Their dynamic natural history was emphasized by Djindjian and Merland (11). Congenital factors, namely the persistence and enlargement of primitive dural arteriovenous communications that normally involute during development, are thought by some authors to be causal (26, 39). The occurrence of dAVMs during childhood, however, is rare. When they do occur in children, these lesions tend to be complex and bilateral, occur more often in male patients, and are associated with cardiac failure and a high mortality rate (38%) (26). Furthermore, most dAVMs occur in middle-aged patients. However, this does not exclude a certain degree of embryological contribution to their pathogenesis, as was shown by Mullan et al. (28). Some patients have had "spontaneous" transverse/sigmoid sinus dAVMs (tss dAVMs) without compromise of the sinus flow (4, 31, 32, 34, 41). Nonetheless, acquired factors are thought to predominate in the cause of tss dAVMs.

Many authors have noted that some degree of flow compromise in the transverse/sigmoid sinus, such as thrombosis, trauma (cranial fracture, craniotomy), infection, previous tumor resection in the area, a hypercoagulable state, pregnancy, hormonal disease, the rupture of an aneurysm, and arterial dysplasia, is associated with tss dAVMs (3, 6, 8, 13, 15, 16, 19, 21, 25, 29, 35, 37). Mironov (23) reported a 72% rate of sinus thrombosis concomitant with tss dAVMs. Fernand et al. (12) observed frank anomalies of venous drainage on the angiograms of all but four patients with tss dAVMs in their series. In these four, the lateral venous sinus appeared to be normal but was associated with opacification of adjacent venous structures. Reports of bilateral sinus occlusion associated with tss dAVMs have also been published (2, 15). A grading system based on the restrictive state of venous drainage in patients with dAVMs has been reported to be useful from a therapeutic standpoint (7, 9, 18, 32). Multiple dAVMs (combinations of cavernous sinus and tss dAVMs or sagittal and tss dAVMs) have also been reported (17, 27).

**Primary sinus compromise**

The prevailing thought is that compromise of the transverse/sigmoid sinus is a primary event that subsequently causes dAVMs. Several authors (16, 22, 37) suggested that dAVMs are an acquired abnormality evolving from revascularization of the previously thrombosed sinus. Other investigators (7, 8, 14, 21) have supported this hypothesis. Awad et al. (3), in their comprehensive meta-analysis, outlined three possible stages in the natural history of dAVMs: 1) sinus thrombosis with engorged dural venous collaterals and the opening of embryonic arteriovenous communications; 2) arteriovenous shunting, which favors the recruitment of arterial feeders into the nidus with secondary venous hypertension; and 3) leptomeningeal retrograde venous drainage, with possible subsequent varicose and aneurysmal dilatation. Bederson (5) hypothesized that the venous hypertension in patients with dAVMs is based on two factors: 1) the increased blood flow through the draining vein caused by a direct shunt into this vein; and 2) the restricted venous outflow, which arises distal to the dAVMs because of the increased blood flow, elevated pressure, and turbulence in the draining vein. According to Bederson (5), these stresses combine to restrict the venous outflow and, in turn, decrease cerebral compliance, elevate intracranial pressure, and even cause hydrocephalus in some patients. Terada et al. (38) induced experimental dAVMs in rats by creating the venous hypertension. They postulated four reasons for this induction: 1) transmission of elevated venous pressure from the dural sinus retrograde to capillaries and arterioles, their resulting dilatation, and loss of sphincter control function; 2) stimulation of angiogenesis, subsequent thickening of vessel walls, and new vessel formation; 3) tissue hypoxia stimulating angiogenesis and angiogenic factor formation; and 4) opening of preexisting microscopic arteriovenous communications.

Another factor reported to compromise transverse/sigmoid sinus flow and precede the generation of dAVMs is previous surgery in the area. Sasaki et al. (36) reported the case of a patient with bilateral tss dAVMs occurring 2 years after the resection of a trigeminal neuroma through a transpetrosal approach. These authors thought that postoperative sinus thrombosis and the apposition of muscle blood vessels to the dura caused the ipsilateral dAVMs and the subsequent elevation of venous pressure in the contralateral sinus. Among 12 cases of tss dAVMs occurring 5 months to 6 years after intracranial surgery (6, 24, 25, 29, 35, 36, 42), 9 were at the site or in the neighborhood of the craniotomy (25, 29, 35, 36). The other three were distant from the original lesion and the craniotomy (6, 24, 42). Five of these 12 lesions developed 2 to 6 years after the sacrifice of the sigmoid sinus, which was involved by a tumor (35). The angiograms obtained before and immediately after the tumor surgery showed no dAVMs. Two of the 12 lesions developed at the site of previous suboccipital craniectomies 1 and 2.5 years later, respectively. These fistulae, according to the authors (29), could have developed after apposition of the vessels of the scalp or muscles to the dura during the initial surgery. Surprisingly, in reported cases of dAVMs associated with transverse/sigmoid sinus compromise, the dominance or nondominance of the compromised sinus is not documented.

**Our experience**

The unique finding in our cases was the association of the tumor invading and occluding the transverse/sigmoid dural venous sinus and the dAVMs. The only other reported case of such association was that presented by Yokota et al. (40), with the coexistence of a meningioma at the transverse/sigmoid sinus, adjacent tss dAVMs, and thrombosis of the ipsilateral sigmoid sinus. Our cases differed in that the sinuses were invaded and occluded by tumors. Nonetheless, our cases further support the hypothesis of an acquired origin of dAVMs. In addition, our cases lend credence to the "causality" hypothesis that the sinus occlusion preceded the formation of the dAVMs (40); this hypothesis was challenged in the commentary to that article. It was postulated that the association of the tumor and the dAVMs might have occurred randomly (10).

Our cases add tumor occlusion of the dural venous sinus to the list of causes
compromising the transverse/sigmoid sinus, which plays a further role in the formation of these dAVMs. Another unique finding was the association of the paraganglioma and the dAVMs, this has not been previously documented. The occlusion of the transverse/sigmoid sinus by two different tumors (meningioma and paraganglioma) adds more weight to the opinion that the tumor is a "nonspecific agent" occluding the sinus and leading to the formation of dAVMs than to the opinion that the tumor induced the formation of the dAVMs by its growth into the sinus (40) or by producing some unknown angiogenic factor. It may also be speculated that highly vascular tumors (meningioma and paraganglioma) might have contributed to the formation of dAVMs by arteriovenous shunting through the highly vascular tumor bed into the dural sinus, elevating its intraluminal pressure.

The dominant venous sinus was invaded and occluded in each of our patients with associated dAVMs. This suggests that the dAVMs developed after the tumor occluded the dominant sinus. We hypothesize that the contralateral, patent, and nondominant venous sinus was not able to manage the additional burden of venous drainage from the occluded side. This factor, we think, combined with the sinus occlusion, was a major contributor to the development of the dAVMs. According to Newton and Potts (30), in only 50% of the population, the drainage from the superior sagittal sinus is primarily or entirely to one (dominant) transverse/sigmoid sinus. This may explain why sinus occlusion is a common clinical event but is relatively infrequently accompanied by dAVMs. Association of the dAVMs can thus be expected in only 25 to 50% of the cases with transverse/sigmoid sinus occlusion.

Subsequently, we have encountered two other cases of meningiomas that occluded the ipsilateral nondominant transverse/sigmoid sinus. No dAVMs were associated with either case, which further supports our hypothesis of "dominant sinus occlusion."

Other considerations

The size, extension, and character of the tumor in each of our patients with dAVMs seemed to determine the extent of the patient's symptoms and whether the symptoms were related predominantly to the tumor or the dAVMs. The symptoms in our first patient (headaches, tinnitus, and objective bruit) related predominantly to the dAVMs; in the second patient (ipsilateral VIIIth-XIIth cranial nerve palsies, contralateral hemiparesis), the symptoms were related to the tumor. The size and extension of these dAVMs, as well as their arterial recruitment, were directly proportional to the size and extension of the tumors. In other words, the larger tumor was associated with the larger dAVMs. In both patients, the tumors were supplied by ipsilateral arteries. The vascularization of both these dAVMs, however, was bilateral. Finally, the blood supply to each tumor was independent of the supply to the dAVMs.

For both patients, treatment consisted of partial preoperative transarterial embolization (to significantly reduce the blood supply to the lesions) and then surgical removal. Early devascularization of the tumor and the dAVMs, respectively, further decreased blood loss and allowed simultaneous excision of the lesions. Any dura or bone involved by either lesion was radically excised.

CONCLUSION

Our experience with two cases of these dAVMs associated with tumors that occluded the transverse and sigmoid sinus supports the hypothesis that dAVMs are acquired and induced. In addition, these lesions were associated with compromise of the flow through the dominant transverse/sigmoid sinus and, supposedly, the inability of the patent, nondominant, contralateral sinus to handle the burden of venous drainage from the occluded side. Finally, our cases add tumor occlusion of the transverse/sigmoid sinus to the list of factors that compromise the sinus and play a role in the cause and pathogenesis of the formation of these dAVMs. The possibility of associated dAVMs should be considered in the diagnostic evaluation of tumors arising adjacent to the dominant transverse/sigmoid sinus.

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Reprint requests: Ossama Al-Mefty, M.D., F.A.C.S., Professor and Chairman, Department of Neurosurgery, University of Arkansas for Medical Sciences, 4301 West Markham, Slot 507, Little Rock, AR 72205-7199.

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This article provides a good description of two cases of dominant sinus occlusion by tumors associated with dural arteriovenous malformations (dAVMs). The association between dAVMs and sinus thrombosis is well recognized, but a clear cause and effect has not been proven. Generally, this association has been noted with spontaneous sinus thrombosis; this report is one of the first showing dAVMs associated with sinus occlusion from a tumor. However, it does not prove that there is a cause and effect. A recent article in Neurosurgery demonstrates that the fistulous connections are within the dura, rather than the sinus (I). The authors of that article suggest, as have others, that the sinus occlusion is an effect of the dAVMs. This and previous reports do not really explain why the arteriovenous malformations are in the dura, and it is possible that the relationship between sinus occlusion and dAVMs is coincidental in most cases. It is important that these observations are clearly described and published, as with this article, so that more knowledge is gained regarding this fascinating disease.

Stanley L. Barnwell
Portland, Oregon

Arnutovic et al. describe two patients who presented with tumors and dAVMs of the transverse and sigmoid sinuses, which they thought were caused by the tumor growth invading and eventually occluding a sinus. I think their observations support the concept that these are acquired lesions that may be related to problems of venous outflow. I do not necessarily think that it must always be the dominant sinus; I have observed dAVMs involving the nondominant transverse/sigmoid sinus, even when tumor is not the potential cause. I do think it is important that patients who undergo the surgical excision of dAVMs are treated with endovascular therapy. They should also undergo follow-up angiography, particularly because these lesions are dynamic. If there are any remaining dAVMs and the outflow has been removed, it is possible that cortical venous drainage can occur as a consequence, with potentially serious implications.