Dural arteriovenous fistulas masquerading as dural sinus thrombosis

Report of 2 cases

Scott Simon, M.D.,¹ Tom Yao, M.D.,¹ Arthur J. Ulm, M.D.,² Benjamin P. Rosenbaum, B.S.,¹ and Robert A. Mericle, M.D.¹

¹Department of Neurosurgery, Vanderbilt University School of Medicine, Nashville, Tennessee; and ²Department of Neurosurgery, Mercer University School of Medicine, Macon, Georgia

The authors report dural sinus thrombosis diagnosed in 2 patients based on noninvasive imaging results, which were revealed to be dural arteriovenous fistulas (DAVs) diagnosed using digital subtraction (DS) angiography. The first patient was a 63-year-old man who presented with headaches. Magnetic resonance venography was performed and suggested dural sinus thrombosis of the left transverse sinus and jugular vein. He was administered warfarin anticoagulation therapy but then suffered multiple intracranial hemorrhages. A DS angiogram was requested for a possible dural sinus thrombectomy, but the DS angiogram revealed a DAVF. The patient underwent serial liquid embolization with complete obliteration of the DAVF. The second patient, an 11-year-old boy, also presented with headaches and was diagnosed with dural sinus thrombosis on MR imaging. A DS angiogram was also requested for a possible thrombectomy and revealed a DAVF. This patient underwent serial liquid embolization and eventual operative resection. These reports emphasize that different venous flow abnormalities can appear similar on noninvasive imaging and that proper diagnosis is critical to avoid contraindicated therapies. (DOI: 10.3171/2008.7.JNS08253)

Key Words • dural arteriovenous fistula • dural sinus thrombosis • intracranial retrograde cortical venous filling • magnetic resonance artifact • venous hypertension

Intracranial dural sinus thrombosis is an uncommon disease in which there is venous thrombosis of one of the dural venous sinuses and perhaps some associated cortical veins. This disease is a difficult problem that can result in venous hypertension, elevated intracranial pressure, and ICH.¹,¹,⁵,⁶,⁸,¹¹ Typically, the treatment for dural sinus thrombosis is anticoagulation therapy, and in some cases endovascular venous thrombectomy or selective dural sinus thrombolysis.²,³,⁷,¹¹

A DAVF is an acquired abnormal connection between a dural artery and a dural venous sinus. A high-flow DAVF can also cause venous hypertension, elevated intracranial pressure and ICH, especially if it is associated with retrograde cortical venous flow.¹,¹¹ Typically the treatment for a DAVF is embolization or resection of the fistula. Anticoagulation therapy is contraindicated in the presence of some DAVFs, especially when they are associated with retrograde venous flow, because of the risk of ICH. In the past 10 years, there has been 1 reported case of a DAVF that was misdiagnosed as a dural sinus thrombosis.¹⁰ Because the standard treatment of dural sinus thrombosis (anticoagulation therapy) is contraindicated in the presence of a DAVF with retrograde cortical venous flow, it is important to obtain the correct diagnosis and proceed with proper treatment.

Case Reports

Case 1

History and Examination. This patient was a 63-year-old man who initially presented with headaches to a neurologist. The patient was in a low-speed car accident in October 2006 and did not seek immediate medical attention. Two days after the accident he suffered the acute onset of aphasia. Magnetic resonance imaging and venography (Fig. 1A) revealed an area of decreased signal intensity in the left parietooccipital lobe and absent dural venous sinuses that were interpreted as infarction and left transverse sinus and jugular vein thrombosis. The patient began war-
farin treatment and was discharged to rehabilitation therapy. One month later, the patient had a seizure witnessed by someone else. Magnetic resonance imaging/venography at that time revealed the same venous flow abnormality and a new left parietooccipital hemorrhage. At the time of this event, the patient’s international normalized ratio was 2.0. The patient’s anticoagulation therapy was discontinued given this new hemorrhage. Nonetheless, 1 week later, the patient complained of the sudden onset of a severe headache and was discovered to have an acute cerebellar hemorrhage. At this juncture the patient’s physicians believed that the risk of extension of his hemorrhage was outweighed by the need to relieve his presumed venous hypertension, and therefore he began to receive therapeutic enoxaparin. One month later, the patient experienced an episode of confusion and a CT scan revealed a third hemorrhage, this time in the same left parietooccipital region as his previous ICH. Enoxaparin was continued and he was referred to neurosurgery for a possible venous thrombectomy.

Operation and Postoperative Course. Intracranial venography revealed a widely patent venous system, but a concomitant 6-vessel cerebral angiogram revealed a high-flow DAVF from the left occipital artery filling a complex dural arteriovenous malformation of the left transverse sinus (Fig. 1B). There was retrograde cortical venous filling on the surface of the left hemisphere, including retrograde flow into the vein of Labbé. The patient’s anticoagulation therapy was discontinued and his fistula was then embolized over 3 sessions using ethylene vinyl alcohol copolymer (Onyx; eV3, Inc.). Complete angiographic obliteration of his DAVF was achieved and confirmed on a 6-month follow-up angiogram (Fig. 1C). He has not experienced any further hemorrhages.

Case 2

History and Examination. The second patient was an 11-year-old boy who was born prematurely (at 26 weeks) with mild developmental delay. Three months prior to admission the patient began experiencing severe daily headaches, occasionally accompanied by emesis. Migraine headaches were diagnosed in the patient and he was treated with multiple medications, but none provided relief. His headaches became more frequent and severe and he eventually missed school on a weekly basis. The day prior to his admission, the patient’s mother witnessed the patient’s collapse, hit his head on the ground, and begin to shake. The patient was taken to an outside hospital where a CT scan revealed no acute disease, and he was transferred to Vanderbilt University Medical Center for further care.

As an inpatient, the patient received an electroencephalogram, a lumbar puncture, MR imaging/venography of the brain, and a CT angiogram of the brain. The patient had an abnormal electroencephalogram result, indicative of mild encephalopathy. No seizure activity was observed. The lumbar puncture revealed no signs of infection, inflammation, or hemorrhage. The MR imaging/venography demonstrated an absence of normal flow in the superior sagittal sinus, right transverse sinus, right sigmoid sinus, and right jugular bulb. The left transverse sinus was at least partially open and the deep cerebral veins were patent. These findings were interpreted as “extensive collateral vessels providing alternate drainage pathways for the sinus thrombosis” (Fig. 2A), which was further interpreted as evidence of extensive dural sinus thrombosis.

Operation and Postoperative Course. As a result of his radiographic study results, the patient began receiving heparin. The neurosurgical service was consulted to explore the possibility of performing a venous thrombectomy. Our recent experience with the aforementioned patient suggested to us that this patient might benefit from an angiogram. A 6-vessel diagnostic angiogram revealed an extensive DAVF with feeders from bilateral middle meningeal arteries, bilateral occipital arteries, bilateral posterior meningeal arteries, and the right meningohypophysial trunk. There was extensive early venous drainage with extensive retrograde venous filling in the dural sinuses and the cortical veins (Fig. 2B and C). The patient’s anticoagulation therapy was discontinued and he underwent 3 staged embolizations using ethylene vinyl alcohol copolymer.
copolymer (Onyx; eV3, Inc.). The patient and his mother reported a marked decrease in headache frequency and severity after each of the 3 embolizations. These embolizations accomplished ~ 50% obliteration of the complex DAVF. The patient subsequently underwent a craniotomy and resection of all remaining arterial feeders until the lesion was completely obliterated.

Discussion

In a paper evaluating 3D TOF MR imaging for diagnosing a DAVF, Chen and colleagues4 compared MR venography results of 7 patients with DAVFs to their DS angiograms. The authors noted that MR imaging revealed 6 of 7 fistulas and none revealed an occluded sinus. Although MR imaging remains a powerful tool with which to evaluate cerebral venous drainage, use of DS angiography remains the standard of care in both adults and children with a clinical suspicion of DAVF.4,9,10,12

Magnetic resonance imaging/venography and CT angiography can lead to diagnostic confusion, especially given that DAVF and dural sinus thrombosis can present with very similar clinical scenarios. This confusion was demonstrated by Kallmes and associates10 in a case report describing a patient presenting with intracranial hemorrhage. An MR venogram suggested a dural sinus thrombosis, but a DS angiogram revealed a DAVF that later was treated using embolization. The authors stated that they performed a DS angiogram because the MR imaging could not rule out a DAVF.

There are 3 possible explanations why TOF MR venography, the most common imaging sequence used for MR venography and the sequence used in these cases, may be prone to misdiagnosing a DAVF as a dural sinus thrombosis.

The first possible explanation might be because TOF MR venography is calibrated to detect flow directed caudally and nullify flow directed cranially, that is, to augment the signal from venous flow away from the brain and minimize the signal from arterial flow toward the brain. The reversal of normal venous flow would therefore result in signal loss that could be misinterpreted as dural sinus thrombosis.

The other 2 possible explanations were offered by Kallmes et al.10 in 1998. First, slow or in-plane flow can generate a low signal on MR venography that can be misinterpreted as a dural sinus thrombosis. Second, the use of an inferior saturation pulse, designed to augment the signal of venous blood, may contribute to signal loss in a dural venous sinus if there is arterIALIZED blood in a sinus from a high-flow fistula.

With these potential pitfalls in mind, it is sometimes possible to determine whether a lack of a signal, suggesting dural sinus thrombosis, is erroneous by examining other sequences that may lead one to perform a DS angiogram. For example, MR angiography, which detects cranially directed flow, and Gd-enhanced sequences can sometimes demonstrate patency of venous structures when retrograde flow is present, but will not necessarily reveal a fistula.

With regard to the cases in this report, it appeared reasonable that the first patient should have initially been given anticoagulants without further diagnostic testing. Because dural sinus thrombosis can be a cause of DAVF development, the diagnostic confusion can be compounded.12 If this occurred, it is possible that anticoagulation therapy would cure the thrombosis but leave the fistula. If this was the case, then the patient had 2 possible origins of a DAVF, trauma or dural sinus thrombosis, and a DS angiogram could have proved useful after the initial hemorrhage.

The second patient presented 1 month after we had treated the first patient, and therefore, we were acutely aware of both the diagnostic confusion between DAVF and dural sinus thrombosis on MR imaging and the usefulness of DS angiography in these cases. The evidence of a venous abnormality in the presence of a negative hypercoagulability workup warranted an angiogram.

In conclusion, the authors believe that diagnostic angiography should be strongly considered in patients with findings on noninvasive imaging consistent with dural

Fig. 2. Case 2. A: Oblique view of cerebral MR venography. White arrowhead shows a small right transverse sinus filling. Small white arrows show prominent cerebral veins with presumed “extensive collateralization.” Black arrows show the absence of the superior sagittal, left transverse, and sigmoid sinus, leading to a diagnosis of extensive dural sinus thrombosis. B: Lateral projection DS angiography sequence of a right ECA injection prior to treatment. Large white arrowhead shows the DAVF as it feeds into the right transverse sinus. Small white arrows show the early appearance of the right transverse sinus, confirming the diagnosis of a DAVF. Small black arrows show the posterior branch of the middle meningeal artery and occipital artery feeders into the DAVF. C: Lateral projection DS angiography sequence of a right vertebral artery injection prior to treatment. White arrowhead shows the DAVF as it feeds into the right transverse sinus. White arrow indicates the early appearance of the right transverse sinus, confirming the diagnosis of a DAVF. Small black arrows demonstrate various posterior circulation arterial feeders into the DAVF.
Two DAVFs masquerading as dural sinus thrombosis

sinus thrombosis prior to initiation of anticoagulation therapy.

Disclosure

Dr. Robert Mericle is a consultant for eV3 Neuroendovascular, maker of Onyx, the liquid embolic agent used in both patients. He is also a consultant for Cordis Endovascular, maker of TruFill, a liquid embolic competitor.

References