Treatment of cranial dural arteriovenous fistulae by interruption of leptomeningeal venous drainage

B. GREGORY THOMPSON, M.D., JOHN L. DOPPMAN, M.D., AND EDWARD H. OLDFIELD, M.D.

Surgical Neurology Branch, National Institute of Neurological Disorders and Stroke, and Department of Diagnostic Radiology, National Institutes of Health, Bethesda, Maryland

Cranial dural arteriovenous fistulae (AVF's) of the tentorial incisura or the dura of the middle fossa have a much higher incidence of draining via leptomeningeal veins than do AVF's of the transverse-sigmoid sinuses or the cavernous sinus. Such a drainage pattern is associated with an increased incidence of intracranial hemorrhage and progressive focal neurological deficits. Patients with cranial dural AVF's often undergo surgical excision and/or endovascular embolization for elimination of the AVF. Since these lesions are frequently large and involve the skull base or adjacent dural sinuses, extensive surgery is often required. In contrast, spinal dural AVF's with only intradural venous drainage to the medullary venous system are treated successfully by simply interrupting the vein that drains the dural AVF as it enters the subarachnoid space. The authors identified a subgroup of patients with cranial dural AVF's in whom the AVF was drained only by leptomeningeal veins, and sought to establish whether simple interruption of the vein draining the blood from the AVF into the subarachnoid space is effective and lasting treatment in this subgroup of patients, as it is in patients with spinal dural AVF's.

Four adult patients with symptomatic cranial dural AVF's (two petrotentorial, one middle fossa floor, and one posterior fossa base) were identified on arteriography as having fistulae that were supplied by the internal and/or external carotid arteries and drained only via leptomeningeal veins (two entered the petrosal vein, one a cerebellar hemispheric vein, and one a mesencephalic vein). All patients underwent interruption of the vein draining the dural AVF as it penetrated the dura to enter the subarachnoid space, and experienced neurological improvement after surgery. Repeat arteriography at 1 to 2 weeks (three patients), 3 months (3 patients), 12 to 15 months (three patients), and 4 years (two patients) revealed no residual AVF and no evidence of abnormal blood flow.

Many cranial dural AVF's with leptomeningeal venous drainage (the type with the most aggressive behavior) are drained only by leptomeningeal veins. This subgroup of patients can be identified by selective arteriography and requires only interruption of the draining vein as it enters the subarachnoid space for successful, lasting elimination.

KEY WORDS • cranial dural arteriovenous malformation • dural arteriovenous fistula • leptomeningeal venous drainage

Cranial dural arteriovenous malformations (AVM's) are arteriovenous shunts imbedded in the dura mater. The location of the AVM nidus in the dura differentiates cranial dural arteriovenous fistulae (AVF's) from the more common parenchymal or pial AVM's. Since 1931, over 400 patients with cranial dural AVF's have been reported and, in large series of intracranial AVM's, cranial dural AVF's account for 10% to 15%. The number of cranial dural AVM's occurring in the United States has been estimated to be between 28,000 and 42,000 annually. Cranial dural AVF's recruit arterial supply from dural arteries and dural branches of cerebral arteries. Extensive fistulae may cause retrograde flow in the dural sinuses and produce increased intracranial pressure. The flow through a dural AVF may also empty into the leptomeningeal veins and cause venous tortuosity, variceal ("aneurysmal") dilatation of pial veins, local or regional intraparenchymal venous congestion, and focal neurological deficits. Although the involved pachymeningeal arteries and veins may be massively dilated, the nidus of the AVF is confined to a localized region of dura.

Treatment options for symptomatic cranial dural
AVF's include surgery, endovascular occlusion, and radiotherapy. Conventional external beam radiotherapy in doses of 15 to 51 Gy has been used to treat cranial dural AVF's, but it takes months to years for thrombosis to occur and the incidence of effective obliteration of the AVF is unknown. Thus, even when irradiation is effective, it has the attendant risk of hemorrhage in the interval between treatment and occlusion, as well as the risk of delayed radiation-induced damage. Stereotactic radiosurgery has not proved beneficial for cranial dural AVF's.

Transarterial and transvenous attempts at endovascular occlusion have occasionally been curative, but are more commonly associated with temporal occlusion only and are generally used as surgical adjuncts. Because the goal of treatment of cranial dural AVF's is elimination of the dural nidus, craniotomy with extensive exposure of the dural sinus and wide dural excision has been recommended. Because the goal of treatment of cranial dural AVF's is elimination of the dural nidus, craniotomy with extensive exposure of the dural sinus and wide dural excision has been recommended.

In contrast to the extensive surgery required to excise the nidus of a cranial dural AVF, dural AVF's of the spine are effectively treated simply by interruption of the arterialized medullary vein that drains the fistula to the subarachnoid space, or with wide dural excision. The goal of the primary goal of treatment of cranial dural AVF's is elimination of the dural nidus, craniotomy with extensive exposure of the dural sinus and wide dural excision has been recommended.

We investigated four patients with cranial dural AVF's, three of whom were referred to the National Institutes of Health (NIH) for treatment of what was originally thought to be a spinal dural AVF. Cases 1 and 2 were included in a previous report that emphasized the need to opacify the internal carotid artery (ICA) and the external carotid artery (ECA) during the arteriographic investigation of patients who appeared to have spinal AVM's, but in whom spinal arteriography was negative. However, the interval of follow-up evaluation after surgery was limited in that publication.

Illustrative Cases

Case 1

This 68-year-old man was evaluated for "claudication." He had reported bilateral calf pain following exercise for several years. In July, 1986, he noticed numbness in both feet and leg weakness. Over the next 4 months, the patient suffered progressive paraparesis and loss of sensation below the beltline. A magnetic resonance (MR) image was suggestive of a subarachnoid filling defect on the axial aspect of the spinal cord, and a myelogram revealed a serpentine filling defect on the dorsal aspect of the spinal cord. Spinal arteriography, including injection of both subclavian and vertebral arteries, failed to demonstrate abnormal vessels. By early 1984, his legs were notably weaker and he had developed bladder dysfunction. After a transient episode of quadripareisis in 1985, complete spinal arteriography was again negative. Carotid arteriography demonstrated a cranial dural AVF at the medial aspect of the right petrous ridge. A frontotemporal craniotomy with attempted excision of the nidus and feeding vessels was unsuccessful. His lower-extremity strength gradually diminished and, by the time of his referral to the NIH in November, 1986, he was unable to ambulate, even with a walker.

Examination. Examination revealed severe paraparesis with bilateral loss of antigravity function in both lower extremities. The upper extremities were normal and there were no cranial nerve deficits. Pinprick sensation was diminished below the L-1 level. Selective spinal arteriography, including injection of both internal iliac arteries, was performed at all levels below T-6 except the intercostal vessels at T-10, T-11, and L-3, which could not be entered because of atherosclerotic changes. No AVF was identified.

Operations. Due to progressive clinical deterioration, an emergency exploratory laminectomy was performed. Following a T10-L4 laminectomy and dural opening, although arterialized veins of the coronal venous plexus were exposed, the nidus of the AVF could not be identified. After surgery, the patient was paraplegic and had no sensation below L-1. He had no rectal tone and was incontinent of bowel and bladder.

On the 3rd postoperative day, left carotid arteriography revealed a dural AVF at the petrous apex supplied by the occipital branch of the left ECA and by the tentorial branch of the ICA. Venous drainage from the dural fistula was solely to the leptomeningeal space via the left petrosal vein to a lateral anastomotic pontine vein and through the foramen magnum to the coronal venous plexus of the spinal cord. A left suboccipital and retromastoid craniectomy revealed a dilated, arterialized petrosal vein, which was ligated, cauterized, and divided as it entered the subarachnoid space.

Postoperative Course. Three months after surgery, the patient had increased leg strength and had regained proprioceptive sense in his feet, but remained unable to walk.

Case 2

This 43-year-old man developed numbness of the left arm, trunk, and leg associated with spasticity of the lower extremities in 1983. A myelogram revealed cervical cord enlargement and a serpentine filling defect on the dorsal surface of the spinal cord. Spinal arteriography, including injection of both subclavian and vertebral arteries, failed to demonstrate abnormal vessels. By early 1984, his legs were notably weaker and he had developed bladder dysfunction. After a transient episode of quadripareisis in 1985, complete spinal arteriography was again negative. Carotid arteriography demonstrated a cranial dural AVF at the medial aspect of the right petrous ridge. A frontotemporal craniotomy with attempted excision of the nidus and feeding vessels was unsuccessful. His lower-extremity strength gradually diminished and, by the time of his referral to the NIH in November, 1986, he was unable to ambulate, even with a walker.

Examination. On admission, the patient was quadriparetic, with more severe weakness and spasticity of the lower than the upper extremities. There was loss of pinprick, vibration, and position sensation in both legs. Selective arteriography delineated a dural AVF of the right medial petrosal apex supplied by the tentorial branch of the right ICA and from the right ECA via the posterior meningeal branch of the occipital artery.
Cranial dural arteriovenous fistulae

(Fig. 1). The right petrosal vein, the sole route of drainage of the fistula, gave rise to retrograde venous flow through the anterior pontomedullary veins inferiorly to enter the coronal venous plexus on the dorsal surface of the spinal cord and through lateral pontine veins to enter a cerebellar hemispheric vein (Fig. 1).

Operation. Endovascular embolization of the fistula with 300- to 500-μm polyvinyl alcohol (Ivalon) particles resulted in arteriographic occlusion of the fistula via the occipital artery, but no clinical improvement. Arteriography performed 3 months later revealed persistent filling of the AVF via the tentorial artery. A right suboccipital craniectomy revealed a dilated, thick-walled petrosal vein conveying arterialized blood from the dura to the subarachnoid space. The petrosal vein was ligated, cauterized, and divided as it entered the subarachnoid space.

Postoperative Course. The postoperative course was marked by gradual, steady improvement. Within 3 weeks, spasticity had diminished and bladder sensation was enhanced. By 3 months, the patient could ambulate with a walker and, at 1 year, he could walk short distances unaided. Selective cerebral arteriography performed at 1 week, 3 months, 14 months, and 4 years after surgery failed to demonstrate residual cranial dural AVF or abnormal flow. At 5 years after surgery, his condition is stable and he has had no evidence of recurrent neurological difficulty.

Case 3

This 44-year-old man developed left-sided headache and orbital pain associated with episodes of nausea and vomiting, without neck stiffness or loss of consciousness, in June, 1987. He also reported bilateral tinnitus which predated the onset of his headaches by several years. In September, 1987, cerebral arteriography revealed a left parasellar dural AVF and he was referred to the NIH.
formed to expose and divide the intrathecal draining
coagulated and divided at the point where the vein
crossed from the dura to the subarachnoid space. A por­
tion of the AVF was cauterized in its dural bed.

Both branches of a single arterialized vein were
coagulated and divided as it penetrated the dura
(large arrow). At surgery, the vein draining the AVF was interrupted as
it penetrated the dura, and the abnormal dural vessels were cau­
tered. Venous drainage of the AVF was via a vein that, after
entering the subarachnoid space (large arrow), bifurcated into
the lateral mesencephalic portion of the left basilar vein and
a left superficial cerebellar vein (not shown). At surgery, the
vein draining the AVF was interrupted as it penetrated the dura
(large arrow). Multiple subsequent arteriograms, obtained as
long as 4 years after surgery and including common carotid
and internal and external carotid injections, failed to opacify
residual AVF or abnormal venous drainage.

Examination. Neurological examination in January,
1988, revealed mild left ptosis, but full excursion of
extraocular muscle movements. There was no audible
cranial bruit. Selective cerebral arteriography revealed
a parasellar dural AVF extending along the posterior
aspect of the lateral wall of the cavernous sinus, the
floor of the medial aspect of the middle fossa, and the
anteromedial portion of the tentorium on the left side.
Venous drainage of the AVF was via a vein that, after
entering the subarachnoid space (large arrow), bifurcated into
the lateral mesencephalic portion of the left basilar vein and
a left superficial cerebellar vein (not shown). At surgery, the
vein draining the AVF was interrupted as it penetrated the dura
(large arrow). Multiple subsequent arteriograms, obtained as
long as 4 years after surgery and including common carotid
and internal and external carotid injections, failed to opacify
residual AVF or abnormal venous drainage.

Operation. A left temporal craniotomy was per­
formed to expose and divide the intrathecal draining
vein. Both branches of a single arterialized vein were
coagulated and divided at the point where the vein
crossed from the dura to the subarachnoid space. A por­
tion of the AVF was cauterized in its dural bed.

Postoperative Course. Postoperatively, the patient
had a transient fourth cranial nerve paresis which dis­
appeared by 6 weeks after surgery. His headaches,
nausea, vomiting, and tinnitus resolved completely.
Arteriography at 2 weeks, 3 months, 15 months, and 4½
years after surgery revealed no residual nidus or abnor­
mal flow. He remains fit and clinically stable 4½ years
after surgery.

Case 4

This 48-year-old man presented to his local physi­
cian in December, 1989, with numbness of his feet.
Two months later, he developed tingling paresthesias
in both legs. In May, 1990, he noted weakness in his
legs, a "stiff gait," urinary incontinence, and altered peri­
adinal sensation. He denied back pain or loss of strength
in the upper extremities.

An MR image of the entire spine was described as
"suggestive of a cervical and a thoracic disc herniation"
and having "abnormal vessels on the posterior aspect
of the cervical and thoracic cord." A myelogram and
postmyelography computerized tomography scan were
"strongly suggestive" of a spinal cord vascular mal­
formation. An MR image of the head was normal. Spi­
nal arteriography was negative. Cerebral arteriography,
including bilateral ICA, ECA, and vertebral artery in­
jections, also failed to demonstrate the nidus of an AVF.
He was referred to the NIH for further evaluation.

Examination. Neurological examination revealed
normal strength, sensation, and reflexes in the patient’s
upper extremities. Examination of the lower extremi­
ties revealed mild weakness and a spastic gait. Muscle
stretch reflexes were markedly increased at both knees
and ankles with sustained clonus. Sensory examination
revealed bilateral diminished position sense of the toes
and hypalgesia and hypesthesia below L-1. A Babinski
response was present bilaterally.

Arteriography of the right common carotid artery re­
vealed an AVF in the dura over the right cerebellar
hemisphere which filled from the meningeal branch of
the right occipital artery (Fig. 3 left). The occipital ar­
tery arose adjacent to the origin of the right ECA, ex­
plaining why the lesion was missed at previous selec­
tive external carotid arteriography. Blood from the
dural fistula drained via a single vein that entered the
intrathecal space to drain into a right cerebellar hemi­
ospheric vein. Retrograde leptomeningeal drainage
conveyed the arterialized flow through the foramen
magnum into the coronal venous plexus of the spinal
cord (Fig. 3 right).

Operation. A right suboccipital craniectomy re­
vealed an engorged, reddish cerebellar hemispheric
vein that crossed the subarachnoid space from the dural
AVF. This vein was coagulated and divided as it left
the dura, and the abnormal dural vessels were cau­
terized.

Postoperative Course. The postoperative course was
marked by rapid improvement. Within 1 week, the pa­
tient began to regain position and vibratory sensation
in the lower extremities. By 3 months, he was able to

FIG. 2. Case 3. Carotid arteriography demonstrating mul­
tiple arteries, including the left external carotid artery, sup­
plying a parasellar dural arteriovenous fistula (AVF, small ar­
wrows) involving the posterior aspect of the lateral wall of the
cavernous sinus, the medial portion of the floor of the middle
fossa, and the anteromedial region of the tentorium on the left
side. Venous drainage of the AVF was via a vein that, after
entering the subarachnoid space (large arrow), bifurcated into
the lateral mesencephalic portion of the left basilar vein and
a left superficial cerebellar vein (not shown). At surgery, the
vein draining the AVF was interrupted as it penetrated the dura
(large arrow). Multiple subsequent arteriograms, obtained as
long as 4 years after surgery and including common carotid
and internal and external carotid injections, failed to opacify
residual AVF or abnormal venous drainage.

B. G. Thompson, J. L. Doppman, and E. H. Oldfield
Cranial dural arteriovenous fistulae

Fig. 3. Case 4. Arteriography of the right common carotid artery (left) and right occipital artery (right) demonstrating filling of a cranial dural arteriovenous fistula (AVF) from a meningeal branch of the occipital artery (left, small arrows). The AVF (right, small arrows), which was located in the dura over the right cerebellar hemisphere, drained into a single vein (left and right, large arrow) that entered the subarachnoid space and conveyed the arterialized blood through the foramen magnum to the coronal venous plexus of the spinal cord. At surgery, the vein draining the AVF was simply interrupted as it penetrated the dura (right, large arrow).

ambulate without a walker. Arteriography at 2 weeks, 3 months, and 14 months after surgery revealed no residual nidus or abnormal flow.

Discussion

Etiology of Symptoms of Cranial Dural AVF’s

Although many authors have suggested, based on individual case reports, that the natural history of cranial dural AVF’s may be related to the pattern of venous drainage, only recently has this relationship been clearly demonstrated by careful review of a sufficiently large number of patients. Two recent studies examined the natural history of cranial dural AVF’s and attempted to identify the risk factors for neurological deficits, hemorrhage, and poor outcome. In a series of 54 patients, 96% of whom were followed for 7 years, reported by Brown, et al., the risk of hemorrhage was 1.5% per year. Significant risk factors for hemorrhage included a location at the petrosal or straight sinus, associated venous varices, and cortical venous drainage. In 1990, Awad, et al., performed a meta-analysis of 377 patients with cranial dural AVF’s previously presented in the medical literature. They identified 100 patients with histories characterized by “aggressive neurological behavior” (hemorrhage or progressive focal neurological deficit other than ophthalmoplegia). Using multiple variant analysis, leptomeningeal venous drainage and “aneurysmal” venous distention were the most significant (p < 0.001) risk factors for aggressive neurological behavior and poor outcome. Patients with leptomeningeal venous drainage were more than 20 times as likely to suffer progressive neurological deterioration or hemorrhage as patients without leptomeningeal drainage. Drainage into the deep (galenic) venous system was also a risk factor (p < 0.05) for aggressive neurological behavior. Awad, et al., concluded that the pattern of venous drainage is the primary determinant of aggressive behavior by cranial dural AVF’s. These findings are also supported by the anatomical findings of other investigators who observed that cranial dural AVF’s bleed through their distended leptomeningeal venous connections, and not from the AVF nidus. The pathophysiology of cranial dural AVF hemorrhage has been explained as follows: arterialized venous drainage produces venous hypertension and leptomeningeal venous drainage which, in turn, leads to variceal or aneurysmal distention and an increased propensity for hemorrhage. Nonhemorrhagic neurological deterioration results from parenchymal venous congestion due to venous hypertension.

Treatment of Cranial Dural AVF’s

The ideal treatment of cranial dural AVF’s should permanently eliminate flow through the AVF and its leptomeningeal venous drainage. In many patients, particularly those with a cranial dural AVF of the transverse sinuses, the AVF is drained by dural veins or by the sinuses, without leptomeningeal venous drainage. Because of the propensity for the development of collateral venous routes to the dural sinuses or to other dural veins, excision of the dural AVF is required for definitive therapy.

In contrast, the patients described here had no drainage of the dural AVF via the sinuses or the dural veins. The fistula was drained solely by veins passing from the AVF into the subarachnoid space via leptomeningeal veins. Since the morbidity rate associated with attempted excision of the dural AVF nidus may be high and because the crucial determinant for poor outcome in patients with cranial dural AVF’s is the presence of leptomeningeal venous drainage, an alternative, less risky, and potentially equally effective treatment would be one that eliminates the venous drainage, permanently eliminates the flow through the dural AVF, and avoids the morbidity associated with excision of the dural nidus. Grisoli, et al., described four patients with tentorial AVF’s who were treated by interrupting the leptomeningeal vein as it left the AVF. At reassessment at 6 months (three patients) and 2 years (one patient) after surgery, there was no evidence of recurrent symptoms; arteriographic re-evaluation at 6 months (two patients) revealed no evidence of flow through the AVF. However, Grisoli, et al., treated only AVF’s of the tentorium, and they did not acknowledge that this approach to treatment depends on the absence of other (dural) pathways of venous drainage. In addition, the arteriographic follow-up examination of their patients was too brief to establish whether recanalization of the AVF occurred after 6 months. In the patients described here,
simple interruption of the intrathecal venous drainage from a cranial dural AVF provided effective, lasting elimination of the clinical and radiographic signs of the lesion and a low morbidity rate and less extensive surgery.

The physiological basis for this simple approach to treatment of the specific subset of cranial dural AVF's described here is supported by the analogous treatment of spinal dural AVF's. Effective treatment of patients with spinal dural AVF's may be accomplished either by excision of the dural nidus or by simple interruption of the medullary vein draining the fistula as it enters the subarachnoid space. In these patients, disconnection of the spinal dural AVF from its intrathecal leptomeningeal drainage permits resolution of venous congestion of the cord and improvement of myelopathy.

Conclusions

Cranial dural AVF's are acquired lesions that develop at certain predictable sites. Fistulae that arise contiguous with the transverse or sigmoid sinuses and lesions of the wall of the cavernous sinus usually produce symptoms due to the rapid flow through them. In contrast, cranial AVF's of the tentorium at the petrous apex and of the floor of the middle fossa usually drain into the leptomeningeal veins and produce neurological symptoms of venous congestion or hemorrhage. The presence of leptomeningeal venous drainage in patients with cranial dural AVF's confers a markedly higher risk of hemorrhage and neurological deterioration. Patients require only interruption of the draining vein as it enters the subarachnoid space for successful lasting elimination of flow through the cranial dural AVF. The fraction of patients who have cranial dural AVF's with only leptomeningeal venous drainage who are amenable to such therapy has not been established.

References


B. G. Thompson, J. L. Dopman, and E. H. Oldfield
Cranial dural arteriovenous fistulae


Manuscript received November 6, 1992.
Accepted in final form February 17, 1993.

Address reprint requests to: Edward H. Oldfield, M.D., Surgical Neurology Branch, National Institute of Neurological Disorders and Stroke, Building 10, Room 5D-37, National Institutes of Health, Bethesda, Maryland 20892.