
R J Ernst, M Gaskill-Shipley, T A Tomsick, L C Hall, J M Tew, Jr and H S Yeh

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Cervical Myelopathy Associated with Intracranial Dural Arteriovenous Fistula: MR Findings Before and After Treatment

Robert J. Ernst, Mary Gaskill-Shipley, Thomas A. Tomsick, Larry C. Hall, John M. Tew, Jr, and Hwa-Shain Yeh

Summary: The MR findings in three patients with intracranial dural arteriovenous fistula associated with cervical myelopathy are described. The MR appearance of an enlarged cord with associated abnormal signal and enhancement is nonspecific and can simulate tumor, demyelination, and inflammation. Enlarged perimedullary vessels may not always be identifiable, but if present, should suggest the presence of an arteriovenous fistula.

Index terms: Fistula, arteriovenous; Spinal cord, myelopathy

Intracranial dural arteriovenous fistulas (DAVFs) are well-known entities that result from an abnormal communication between meningeal arteries and veins and/or venous sinuses. Intracranial DAVFs can produce a variety of clinical manifestations, including intracranial hemorrhage, ischemia/infarction, mass effect from enlarged veins, increased intracranial pressure, and cranial neuropathies. Myelopathy resulting from an intracranial DAVF is uncommon. Recognition of the magnetic resonance (MR) imaging findings is important, since many of the imaging characteristics are nonspecific. If not properly identified, a delay in diagnosis can result in progression of the myelopathy or in an unwarranted cord biopsy.

We report the MR findings in three patients in whom progressive cervical myelopathy developed as a result of intracranial DAVF. Of the 25 cases we found previously reported in the literature, the MR features were detailed in eight cases. MR findings in these eight patients ranged from normal to cord enlargement with associated signal abnormality of the cord and enlarged perimedullary vessels (1–6). We present the MR features related to this myelopathy before and after endovascular and surgical therapy and discuss the importance of identifying enlarged veins adjacent to the spinal cord abnormality and of following patients after treatment.

Case Reports

Case 1

A 71-year-old man was admitted with progressive nausea and vomiting and bilateral leg weakness. MR images of the brain and cervical spine revealed abnormal hyperintensity on the long-repetition-time (TR) images in the lower medulla and upper cervical cord to the C-3 level. Enlarged perimedullary vessels were present along the lower brain stem, cervical cord, and upper thoracic cord (Fig 1A–C). Angiography revealed an intracranial DAVF of the superior petrosal sinus with perimedullary venous drainage (Fig 1D and E).

Embolization was not attempted, since the meningeal branches arising from the internal carotid artery were the predominant supply, and their safe catheterization and occlusion were not assured. Surgery was performed to obliterate the fistula. Postoperatively, the extremity weakness improved dramatically. A repeat MR examination 10 days after surgery showed improvement in brain stem signal abnormality and complete resolution of the abnormally dilated veins (Fig 1F). The patient has remained neurologically stable with improvement in leg strength over an 18-month period.

Case 2

A 47-year-old man had a 5-month history of progressive myelopathy. MR imaging showed mild cord enlargement with abnormal hyperintensity on the long-TR images. Subtle signal voids were present ventral and dorsal to the cervical cord (Fig 2A). A DAVF, which was identified medial to the occipital condyle at angiography, was supplied predominately by the right ascending pharyngeal artery (Fig 2B). The fistula was successfully treated by endovascular embolization with 0.1 mL of N-butyl-cyanoacrylate (Fig 2C). After embolization, mild symptomatic
improvement occurred with no further progression of the myelopathy. A 4-year follow-up MR study showed a normal cervical cord without enhancement or abnormally dilated veins (Fig 2D). The thoracic cord had normal signal with mild to moderate cord atrophy.

Case 3

A 58-year-old woman was known to have had an extensive DAVF of the skull base for many years. Previous treatments included attempted local resection, left external carotid artery ligation, left vertebral artery ligation, and external beam radiation. She presented with progressive cervical myelopathy. MR imaging revealed a mildly enlarged cervical cord and hyperintensity within the cord on the long-TR images. Enlarged vessels dorsal and ventral to the cord were subtle but present (Fig 3A and B).

Angiography showed an extensive DAVF of the skull base with arterial supply via collaterals to the occluded external carotid artery from the left ascending cervical, left vertebral, and left ophthalmic arteries (Fig 3C). The dural fistula was so extensive, with limited arterial access, that complete occlusion either via embolization or surgery was not thought to be achievable. Partial embolization was performed to diminish arterial inflow, thereby decreasing venous pressure in the hope of stabilizing the patient’s cervical myelopathy. Embolization was performed using polyvinyl alcohol and silk thread. After embolization, the symptoms improved transiently. Over the ensuing months, her condition worsened with increased spasticity and muscle weakness. A follow-up MR examination at 4 years showed persistent cord signal abnormality, cord swelling, and abnormal cord enhancement; however, no enlarged veins were apparent (Fig 3D). Although no enlarged perimedullary veins were identified, the patient’s clinical course was compatible with a worsening fistula due to recruited collaterals or recanalization.

Discussion

Recognition of the MR findings of DAVFs is important, because most patients with symptoms of myelopathy are initially examined by MR imaging. An improper diagnosis can result in an unwarranted cord biopsy or a delay in
diagnosis with progression of myelopathy. These three cases are unusual because the du-
rnal fistulas were located intracranially with sec-
ondary effects on the cervical cord.

Intracranial, craniocervical, and spinal
DAVFs are uncommon causes of myelopathy.
Myelopathy associated with DAVFs is related to
venous hypertension with subsequent edema,
hypoxia, or both (7). In our three patients, the
site of the fistula was remote from the level of
the spinal cord dysfunction.

Wrobel et al (6) described three patients who
had cranial DAVFs with associated myelopathy
that were documented by angiography. In all
two cases, the DAVFs were primarily supplied
by the carotid circulation. Venous drainage
communicated to the posterior spinal veins via
the petrosal vein. These cases support the the-
ory that neurologic impairment is related to ve-
nous hypertension and not arterial insufficiency.

Unlike parenchymal arteriovenous malfor-
mations, the abnormal arteriovenous commu-
ication in DAVFs is within the dura or the wall
of the dural sinus. The nidus is usually small and
often not identifiable on routine spin-echo MR
images. MR findings of intracranial DAVFs are
often subtle and include enlarged venous struc-
tures, occluded venous sinuses, associated pa-
renchymal edema, infarction, and hemorrhage
(8). These findings usually relate to the diver-
sion of venous flow from the sinuses to the
cerebral veins. Flow-related enhancement
within and enlargement of the venous sinuses
may be present on three-dimensional time-of-
flight images (9).

In our three cases of intracranial DAVF with
associated cervical myelopathy, the MR find-
ings included cervical cord enlargement and
abnormal hyperintensity on the long-TR images
within the medulla and cervical cord. Serpigi-
nous areas of signal void located ventral and
dorsal to the cord representing enlarged peri-
medullary veins were present in all cases. Con-
trast material was administered intravenously in
only one case and produced enhancement of
the enlarged venous plexus. On the basis of
findings in spinal DAVFs, variable parenchymal
and vascular enhancement would not be unex-
The MR findings of an enlarged cord with abnormal signal are nonspecific and can easily be mistaken for tumor, demyelination, or myelitis. The identification of enlarged veins should suggest the presence of a DAVF, which can be confirmed with angiography.

Similar changes—including cord signal abnormality, cord enlargement, and enlarged perimedullary vessels—are seen with spinal DAVFs (11, 12). Spinal DAVFs may produce changes at levels above or below the fistula itself, owing to transmission of venous hypertension via the coronal venous plexus. However, we have not found signal abnormalities of spinal DAVFs reported to extend into the medulla, as in our three cases. The findings of intracranial DAVFs on routine spin-echo MR sequences are subtle and may not be apparent. Therefore, negative findings at spinal angiography performed to search for a spinal DAVF should be followed by cerebral angiography to exclude a cranial DAVF with spinal venous drainage. In addition, sacral DAVFs can occur with ascending venous engorgement and venous hypertension leading to thoracic myelopathy. Therefore, if both the spinal and cerebral arteriograms are negative for the presence of a DAVF, a flush aortogram to exclude a sacral DAVF is indicated.

The abnormal signal within the cord and associated cord enlargement is thought to represent edema from venous hypertension. Associated cord ischemia with demyelination and/or venous infarction may occur, which may account for the persistent neurologic deficit following successful treatment of the fistula. The serpiginous areas of signal void around the cord represent enlarged veins. Demonstrated caudal flow via intracranial-to-spinal venous communications explains the transmission of venous hypertension to the coronal venous plexus of the upper spine. On the basis of MR findings in cases of spinal DAVFs, areas of signal void identifiable on routine spin-echo images would not be expected in all cases of intracranial DAVFs. Bowen et al (13) identified enlarged vessels on routine MR images in five of eight cases of spinal DAVFs. Gilbertson et al (14) identified flow voids on long-TR images in 45% of 44 cases of spinal DAVFs. MR angiography may be beneficial, especially in cases of suspected DAVF in which there are no identifiably enlarged vessels on routine spin-echo images. Bowen et al (13) identified abnormal vessels in all eight of their cases at MR angiography after contrast administration.

On the basis of our findings and on reports in
the literature, the radiologic evaluation of suspected spinal DAVFs should include a routine contrast-enhanced MR examination of the spine. If abnormal signal is present within the cord on long-TR images, and no abnormal veins are identified on the spin-echo images, then MR angiography and myelography are recommended. Angiography is required to confirm the presence of and to characterize the fistulous site.

Analogous to the treatment of spinal DAVFs, the goal of therapy is complete occlusion of the intracranial dural fistula, thereby stabilizing or reversing neurologic symptoms (3, 15). Improvement of the myelopathy varies after treatment depending on the duration of the fistula and the degree of permanent cord damage; however, stabilization of signs and symptoms is usually achieved. Complete reversal of clinical deficits was not achieved in any of our patients despite the apparent cure in cases 1 and 2. Multiple previous surgical procedures led to limited arterial access in patient 3 and prohibited complete obliteration, allowing for subsequent deterioration.

In conclusion, we documented the MR findings in three patients who had intracranial DAVFs with associated cervical myelopathy. Findings included abnormal cervical cord signal extending into the medulla, mild cord enlargement, and the identification of enlarged perimedullary veins. On the basis of findings in cases of spinal DAVFs, abnormally enlarged perimedullary vessels would not be expected in all cases. In such cases, a high degree of clinical suspicion in association with the nonspecific findings of cord swelling and edema is required to suggest the diagnosis of a DAVF. These cases emphasize that DAVFs may be remote from the level of myelopathy, which is important when performing angiography in suspected cases of spinal DAVFs. MR imaging is useful in following up patients after treatment to evaluate cord signal abnormality and dilated veins.

References