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This information is current as of June 6, 2023.

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AJNR Am J Neuroradiol 1998, 19 (3) 583-586
<http://www.ajnr.org/content/19/3/583>

Craniocervical Dural Fistula Associated with Cervical Myelopathy: Angiographic Demonstration of Normal Venous Drainage of the Thoracolumbar Cord Does Not Rule Out Diagnosis

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Summary: We report a case of craniocervical dural arteriovenous fistula with perimedullary venous drainage associated with cervical myelopathy in which spinal angiography showed a normal venous phase after injection of the artery of Adamkiewicz. We conclude that because of the complex venous drainage of the spinal cord, a dural arteriovenous fistula with spinal drainage cannot be ruled out solely because a normal venous phase is seen in the lower part of the cord, as has previously been suggested.

Spinal dural fistulas with perimedullary venous drainage are the most frequently encountered spinal arteriovenous malformations (1-3). By comparison, intracranial dural arteriovenous fistulas (DAVFs) draining into the perimedullary venous system are rare lesions that may be accompanied by symptoms and imaging abnormalities similar to those of the more frequent spinal DAVFs (4-10). Because of the venous congestion of the cord associated with the presence of a DAVF, it has been said that the presence of a normal venous phase of the Adamkiewicz artery injection during angiography should rule out the presence of a dural fistula with perimedullary drainage, even if the fistula is located intracranially (11). We report a case of a craniocervical junction DAVF with perimedullary drainage presenting as a cervical myelopathy in which venous compartmentalization accounted for the isolated abnormal signal of the cervical cord and the normal venous drainage of the lower cord at magnetic resonance (MR) imaging.

Case Report

A 74-year-old man had progressive paraparesis over a period of 1 year. His medical history included prostate cancer treated by prostatectomy and radiation therapy. Physical examination revealed severe paraparesis, accompanied by marked spasticity. There was diffuse hyperreflexia in the four limbs, more marked in the lower extremities. Plantar reflexes were in extension bilaterally. No sensory level could be established. Weakness of the upper limbs, mainly limited to the intrinsic muscles of the hands, was also noted. An MR study of the entire spine showed enlargement of the cervical cord, as well as a diffuse hyperintense intramedullary signal on T2-weighted images, extending from the level of C-2 to approximately T-2 (Fig 1A). The rest of the cord was normal in appearance (Fig

1B). Dilated veins were noted on the dorsal surface of the thoracic cord, down to the level of the thoracolumbar junction (Fig 1C). Degenerative changes were also noted at the cervical level, causing some degree of spinal stenosis.

Selective angiography of both vertebral, subclavian, and external carotid arteries revealed the presence of a dural fistula of the foramen magnum, supplied by a meningeal branch originating from the left vertebral artery, proximal to the origin of the left posterior inferior cerebellar artery (Fig 1D). The fistula drained into a dilated radicular vein, which joined the anterior medullary vein, anastomosing with posterior medullary veins. Venous drainage of the fistula extended down to the level of the lumbar spine, where it shared the same radicular vein draining the territory of the artery of Adamkiewicz to reach the epidural space (Fig 1E). Other minor drainage routes were present via small radicular veins that reached cervical and thoracic epidural veins. The venous drainage of the thoracolumbar cord was normal. Visualization of the draining radicular vein occurred between 10 and 14 seconds after injection of the artery of Adamkiewicz originating from the right ninth intercostal artery (Fig 1F).

At the time of diagnostic arteriography, a superselective injection of the dural fistula was performed, but reflux into the vertebral artery did not allow safe embolization. The patient was thus referred for surgical treatment. Via a left suboccipital craniotomy and after C-1 laminectomy, the large draining vein was exposed and sectioned. The postoperative course showed only partial improvement. Strength was 5/5 in the upper limbs with restored sensory function, but some residual lower limb weakness and spasticity persisted. The patient could ambulate with the use of a walker 3 months after surgery. Controlled angiography performed 2 months after surgery confirmed obliteration of the DAVF. MR imaging of the spine was not performed.

Discussion

Intracranial DAVFs with perimedullary venous drainage are rare lesions. We found 33 cases reported in the English and French literature (4-10, 12-17). These vascular anomalies were labeled type V intracranial DAVFs in the revised classification scheme of Djindjian and Merland (18). Diagnosis may be difficult because they share similar clinical and radiologic

Received January 3, 1997; accepted after revision March 20.

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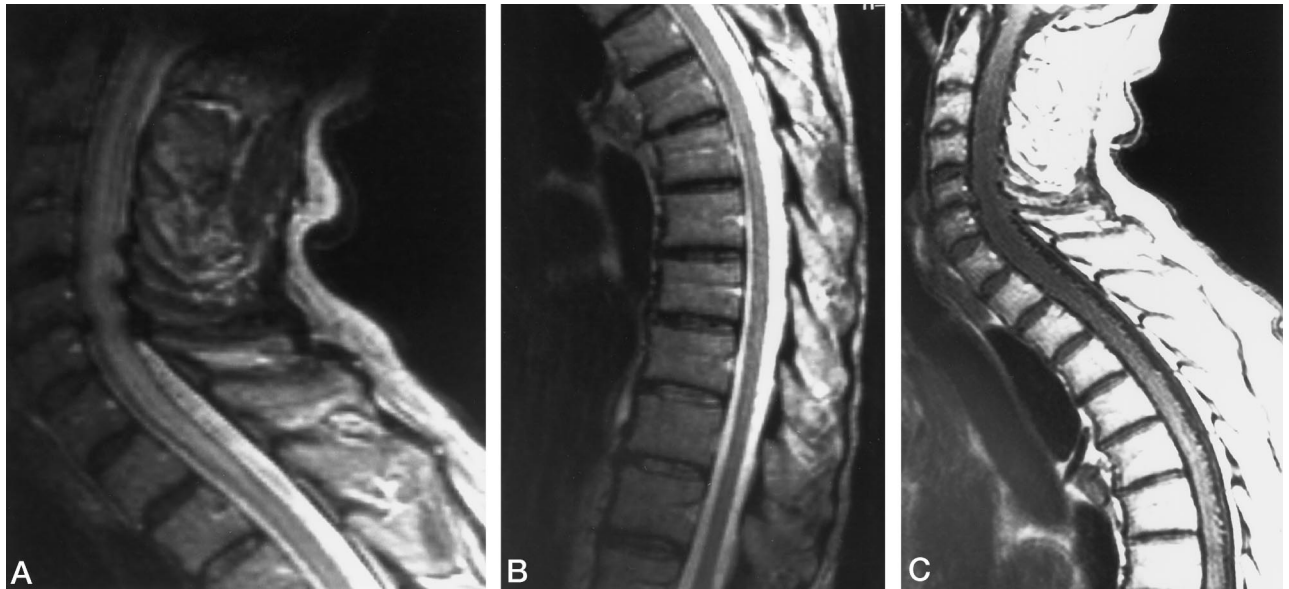


FIG 1. A–C, MR appearance of the cord in a 74-year-old man with severe paraparesis and marked spasticity.

A, T2-weighted image at the cervical level shows enlargement of the cord and increased signal intensity.

B, T2-weighted image at the thoracolumbar level shows normal appearance of the cord.

C, T1-weighted contrast-enhanced image at the thoracolumbar level shows an enlarged vessel on the posterior surface of the cord.

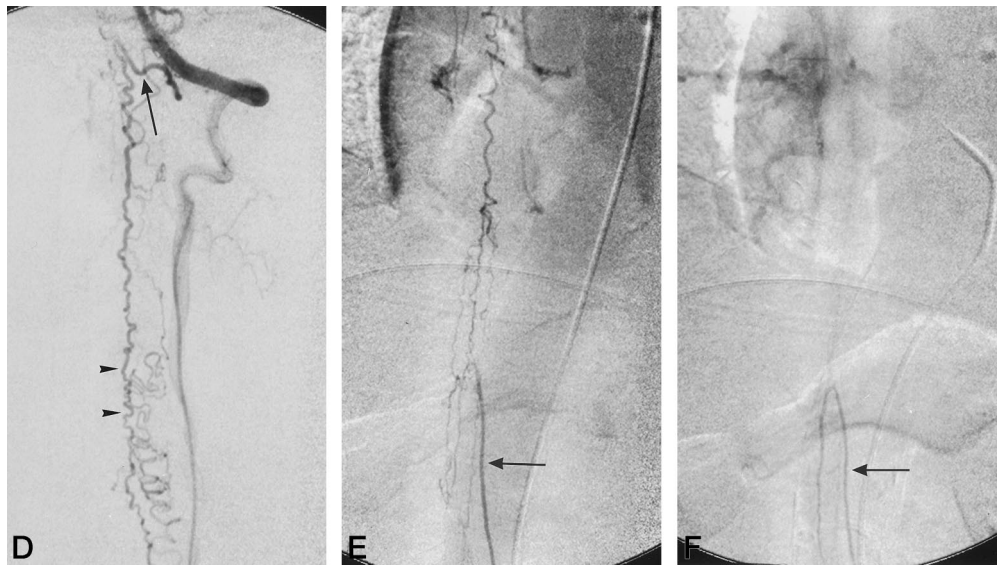
D–F, Findings at left vertebral arteriography.

D, Posteroanterior view at the cervical level shows the DAVF fed by a meningeal branch of the vertebral artery.

Arrow points to the radicular vein; arrowheads point to the congested veins on the surface of the cord.

E Same injection in a later phase centered at the thoracolumbar junction. Note that the draining veins of the fistula reach the radicular vein of L-2 on the left (arrow) to exit in the epidural space.

F, Injection of the left ninth intercostal artery, venous phase at 12 seconds. Note that the radicular vein draining the territory of Adamkiewicz is also the left L-2 radicular vein (arrow).



findings with the much more frequent spinal DAVFs (4–10). In both diseases, the arteriovenous shunt causes a chronic venous congestion of the cord and secondary ischemia (1), which is responsible for the clinical picture of ascending myelopathy. A poorly understood venous disorder is evoked to explain why this low-flow shunt cannot find its way into the epidural plexus (7, 12). Degenerative changes of the spine may contribute to a reduction of the potential reservoir of draining radicular veins. Clinical symptoms include progressive paraparesis, bowel and bladder dysfunction, and sensory abnormalities (1, 19). The symptoms do not relate to the site of the fistula itself, which may be situated anywhere from the tentorium to the sacrum (20).

Classically, the diagnosis is suggested by the myelographic and MR anomalies: dilated veins are seen on the posterior surface of the cord, which usually has an increased signal intensity on T2-weighted images, most often starting at the caudal extremity (2, 20). Discrete or pial surface enhancement after injection of contrast material has also been reported (20). According to the pathophysiology of venous congestion, injection of the major radiculomedullary artery of the thoracolumbar cord (artery of Adamkiewicz) at spinal angiography characteristically shows stasis in the arterial phase and delayed or absent venous drainage (11, 21).

Although MR findings were not available in the first case reports of this condition, the presence of

abnormal intramedullary signals limited to the cervical cord has been well documented in six cases of intracranial DAVFs with perimedullary venous drainage (4–6, 10, 15, 17). Abnormal signals in the lumbar region and the conus medullaris, with varying degrees of cephalad extension, have also been reported (9). In these cases, the congestion is probably transmitted to the lower cord via venous anastomoses. The lower part of the cord may be more vulnerable because of the addition of the effect of gravity.

Normal venous drainage of the cervical cord occurs via lateral epidural plexus or continues cephalad via the perimedullary venous plexus toward the posterior fossa (21). The dorsal level represents a hinge region and drainage of this segment can proceed in a cephalad or a caudal direction while lumbar drainage proceeds caudally or laterally toward the epidural plexus (21). Venous opacification after injection of the Adamkiewicz artery is usually visible at 13 seconds and disappears after 21 seconds (7, 21). Angiography in our patient showed how the enlarged and tortuous medullary veins extend from the upper cervical cord to the level of the lumbar spine, where they join the same radicular vein draining the territory fed by the artery of Adamkiewicz. This anatomic condition was insufficient for drainage of the cervical level, as signs of venous congestion were present. Injection of the artery of Adamkiewicz revealed normal venous drainage, and the MR appearance of the lower cord was normal, suggesting that the DAVF did not interfere with the venous drainage of this part of the cord.

This is an unusual case of a DAVF with medullary venous drainage not associated with an abnormal venous phase at injection of the artery of Adamkiewicz during an active phase of the disease. Several reports on intracranial DAVFs with perimedullary drainage do not mention whether the venous phase of the artery of Adamkiewicz was studied or not (4, 6, 8, 10). However, an abnormal venous phase was present in all the studies in which it was looked for (7, 9, 13, 14). Gobin et al (7) described one case in which a normal venous phase was noted after injection of the artery of Adamkiewicz, but they stressed that this occurred during a period of clinical remission; indeed, when angiography was repeated while the patient was symptomatic, the venous phase was delayed. A similar observation was reported by Mahagne et al (14).

Willinsky et al (11) stated that "if the venous phase of the spinal circulation is normal, this alone rules out DAVF as the cause of the patient's symptoms." When a DAVF with perimedullary drainage is suspected, these authors suggest starting the angiography by finding the Adamkiewicz artery, and that in the presence of a normal venous phase, the angiographic examination could be stopped. In our case, the normal venous drainage time of the lower cord associated with normal signal of the dorsal cord and conus on MR images suggested that there was a hemodynamic compartmentalization between the cervical cord and the lower cord, even if there was an anatomic continuity between both venous systems. Thus, in this case, since only the venous drainage of the

cervical cord was in jeopardy, the normal venous drainage of the artery of Adamkiewicz could have led us to erroneously rule out the presence of a DAVF.

Conclusion

Because of the complexity of the cord's venous system, the presence of an intracranial DAVF with perimedullary drainage can not be ruled out solely on the basis of a normal venous phase of the Adamkiewicz artery injection. When the presence of a DAVF is suspected, and the abnormality of the cord is limited to the cervical level on MR images, we believe that the angiographic protocol should start with posterior fossa angiography and injection of the external carotid artery.

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