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Bilateral Cervical Spinal Dural Arteriovenous Fistulas with Intracranial Venous Drainage Mimicking a Foramen Magnum Dural Arteriovenous Fistula

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Summary

We describe a unique case of bilateral cervical spinal dural arteriovenous fistulas mimicking an intracranial dural arteriovenous fistula near the foramen magnum. We review its detection via MRI and digital subtraction angiography and subsequent management through surgical intervention. Pitfalls in diagnostic angiography are discussed with reference to accurate location of the fistula site. The venous anastomotic connections of the posterior midline spinal vein to the medial posterior medullary vein, posterior fossa bridging veins, and dural venous sinuses of the skull base are discussed with reference to problem-solving in this complex case. The mechanism of myelopathy through venous hypertension produced by spinal dural fistulas is also emphasized.

Introduction

Spinal dural arteriovenous fistulas (SDAVF) are the most common spinal vascular malformation [1,2]. It is unusual to identify more than one SDAVF in a single patient, and even more unusual to demonstrate intracranial venous drainage from a SDAVF unless the fistula site is located near the skull base. Marginal sinus fistulas are a type of intracranial dural arteriovenous fistula (DAVF) that typically derives its arterial supply from branches of the external carotid and vertebral arteries, with rapid drainage to the condylar or internal jugular veins. Both SDAVF and posterior fossa intracranial DAVF can cause myelopathy depending on the pattern of venous drainage causing brainstem or spinal cord congestion [3,4]. We describe a case of bilateral SDAVF arising at the same level in the lower cervical spine, with associated intracranial venous drainage via the posterior midline spinal vein that led to diagnostic confusion, misidentification of the fistula as a posterior fossa intracranial DAVF, and thus an initial surgery in the posterior fossa that was not curative, followed by a second curative surgery in the lower cervical spine.

Materials and Methods

This case report includes radiographic and clinical information in a single patient. Data was analyzed retrospectively in accordance with a protocol approved by the UCSF Committee on Human Research.

Case Report

A 40-year-old man presented with acute quadriparesis after a febrile illness and was found on MRI (Figure 1) to have extensive cer-

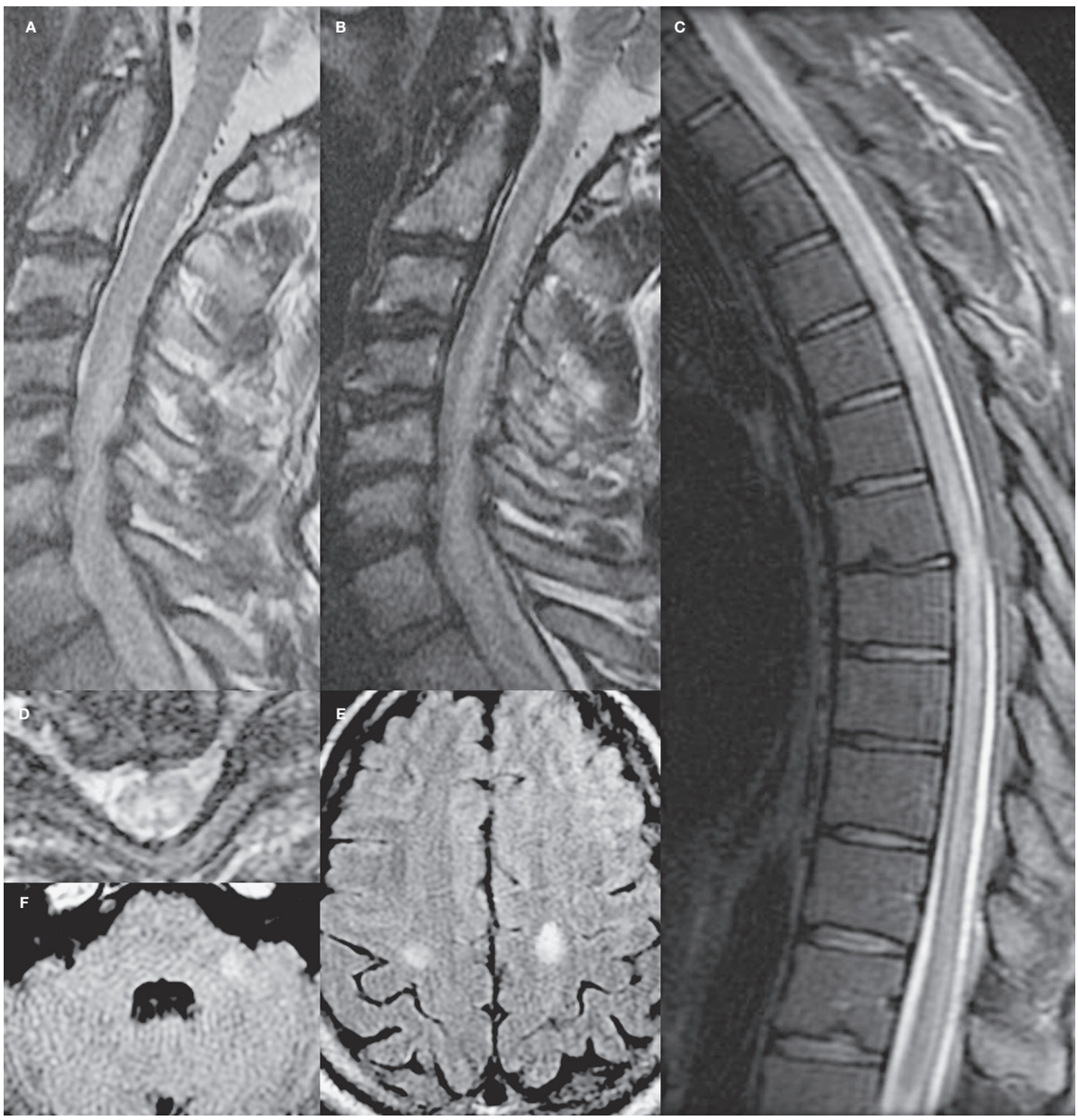

Figure 1 Sagittal and axial T2 weighted MR images of the cervical and thoracic spine at the time of initial presentation demonstrate patchy discontinuous long-segment T2 prolongation in the central cervical and thoracic spinal cord (A-D) with associated enlarged flow voids along the dorsal surface of the cervical spinal cord and cervicomedullary junction (A,B). Degenerative spinal canal stenosis is also evident in the lower cervical spine, severe at C5/6. Axial FLAIR images of the brain demonstrate additional focal T2 hyperintense white matter lesions involving the corticospinal tracts (E) and brachium pontis (F).

vical and thoracic spinal cord edema as well as several posterior fossa and supratentorial white matter lesions. The patient was treated with corticosteroids for a diagnosis of acute disseminated encephalomyelitis. Over several weeks, nearly full motor strength was regained.

Despite improved motor function, the patient developed severe pain in the distal upper and lower extremities and had persistent urinary and bowel retention. A follow-up MRI five months after initial presentation demonstrated resolution of spinal cord edema, but persistent enlargement of an intrathecal vessel dorsal to the cervical spinal cord (Figure 2A). A digital subtraction angiogram (DSA) was then performed (Figure 2B-F).

DSA identified hypertrophied radicular branches of the bilateral costocervical trunks at C5/6 converging on a single enlarged vessel dorsal to the spinal cord. The rapidly shunting dorsal vessel coursed into the posterior fossa, turned laterocaudally and entered the dura just above the foramen magnum, medial to the marginal sinus, and emptied into the epidural venous plexus between the occiput and C1 (Figure 2B,D). Based on these images, it was concluded that the fistula site was at the posterior craniocervical junction at the dural exit site just below the foramen magnum, with the vessel dorsal to the spinal cord being a feeding artery deriving bilateral supply. A suboccipital craniotomy and clipping of the enlarged laterocaudally coursing posterior fossa vessel was performed at its entry point into the dura just above the foramen magnum. Postoperative angiography, however, demonstrated redirection of shunting flow into brainstem veins that then decompressed into the cavernous sinuses (Figure 3). The patient remained intubated and was kept hypotensive overnight due to presumed elevated brainstem venous pressures; he was returned to the operating room the next morning. Based on the results of the second angiogram, the patient was now presumed to have bilateral spinal dural arteriovenous fistulas arising from the right and left radiculomedullary arteries at C5/6, with two separate fistula sites draining to a common vein. The enlarged vessel dorsal to the cervical spinal cord and medulla was thus deemed to represent the posterior midline spinal vein (continuing as the medial posterior medullary vein), that in its cranial end had originally taken a broad turn to the right (via a bridging vein) before exiting the inner leaf of the dura in the posterior fossa and decompressing into the marginal sinus and epidural venous plexus.

A second stage surgery was performed with laminectomy and clipping of bilateral intrathecal veins emanating from bilateral C5/6 SDAVF (with two fistula sites now presumed to be in the C5/6 neural foraminal dura and convergence of right and left radiculomedullary veins on the enlarged posterior midline spinal vein at the same level). DSA following the second stage surgery then confirmed elimination of AV shunting. The patient was extubated and had slow improvement in extremity strength during his hospitalization. Urinary and bowel retention were still present upon discharge, but slowly improved over the following six months. MRI at that time confirmed no residual enlarged intrathecal flow voids and no spinal cord edema (Figure 4).

Discussion

Although multiple SDAVFs have been reported in the same patient, this case of bilateral lower cervical SDAVF arising at the same spinal level and converging on the posterior midline spinal vein at the same point appears to be unique. A pair of mirror-image SDAVF at the craniocervical junction has been reported [5], as have bilateral thoracic fistulas with multiple points of shunting to different intradural veins [6]. Initial diagnostic confusion in this case was due not only to the symmetric bilaterality of cervical fistulas but also to the tortuosity of the pressurized midline posterior spinal vein, thus leading to uncertainty as to whether the structure represented a truly midline vein or instead a hypertrophied posterolateral spinal artery (PLSA). In retrospect, the extreme tortuosity of this structure could have directed us toward identifying the structure as a vein (more prone to becoming tortuous under conditions of high flow) instead of an artery.

On DSA, recognition of the site of arteriovenous shunting is critical in the analysis of any fistula. This is achieved by careful analysis of each frame of the DSA, often using high frame rates to visualize the fistula site. In a spinal dural arteriovenous shunt there is often a transition between the smaller caliber of the meningeal feeder and the larger draining radicular vein. Often magnification and oblique views are helpful in delineating the fistula point, as can be 3D rotational angiography. Although oblique and 3D images were acquired in this case, frame rates in excess of four per second were not achievable on the specific angiography unit used when it was configured in biplane mode. Had only the single AP plane been used at frame rates in excess of eight per second, the DSA might have allowed us to localize the fistulas accurately in the lower cervical spine instead of the posterior fossa.

Diagnostic confusion was also compounded by observation of the laterocaudal course of the intracranial vascular structure connecting the midline dorsal tortuous vessel to the lateral dura above the foramen magnum, mimicking the appearance (though not the direction of flow) of a PLSA (though PLSA should not be present intracranially). Note that the tight hairpin turn

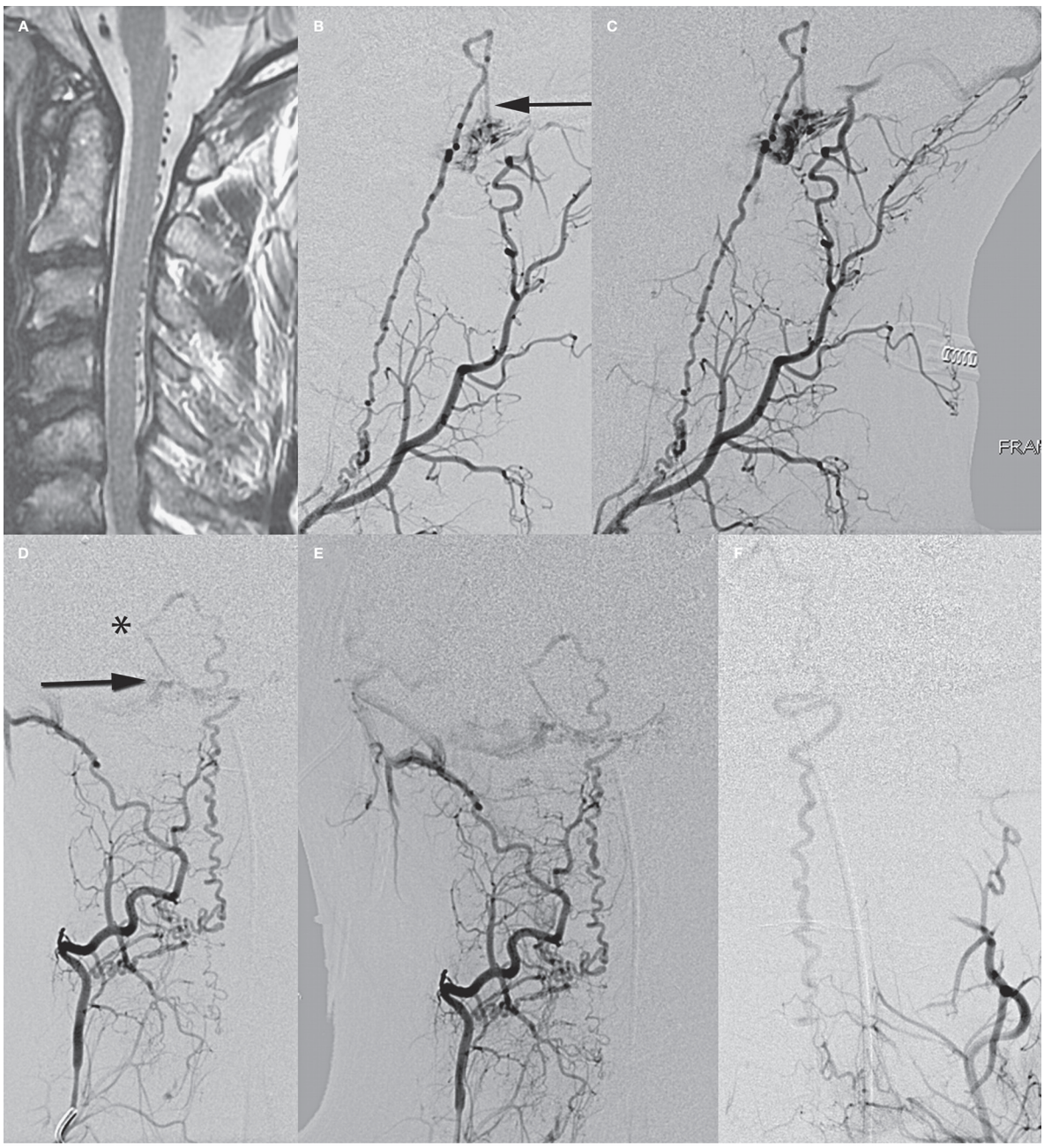

Figure 2 Sagittal T2 MRI 5 months after initial presentation shows resolution of spinal cord edema, but persistence of enlarged flow voids dorsal to the cervical spinal cord and medulla (A). DSA shows right (D,E) and left (F) costocervical artery supply to the enlarged midline intrathecal vessel, confirmed to run dorsal to the spinal cord by injection of the right costocervical artery during lateral DSA (B). Dural entry point of the shunting vessel (*) and presumed fistula site at the exit point to the epidural venous plexus (black arrows) are marked.

of the PLSA (radiculopial artery) is not evident in the cervical spine. Instead, cervical PLSAs are part of a posterior-lateral anastomotic network and therefore should not be seen as a single midline vessel over a long segment of the spine [7]. Both SDAVF and foramen magnum DAVF are classified as lateral epidural DAVFs [1,8]. In SDAVF, the fistula site is located in the neural foramen at the lateral dural egress of the bridging radiculomedullary vein to the epidural venous plexus. The bridging lateral medullary veins serve a similar role as the radiculomedul-

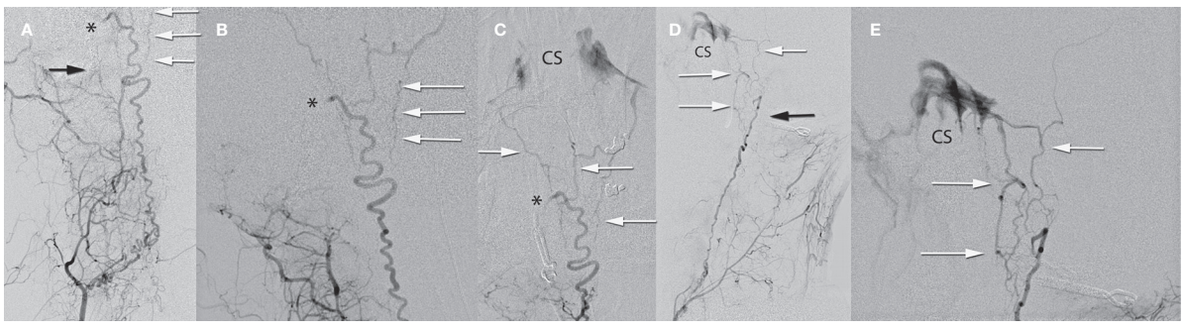

Figure 3 Frontal (A) and lateral (D) DSA of the right costocervical artery following surgical clipping of the intradural vessel in the posterior fossa at its entry point into the dura (*) as well as its exit point to the epidural venous plexus (black arrow). Although there is no longer filling of the epidural venous plexus between the occiput and C1, there has been redirection of blood flow to veins along the surface of the medulla, pons, and midbrain (white arrows) that now decompress into the cavernous sinuses (cs) bilaterally. Similar results were found upon injection of the left costocervical artery (not shown).

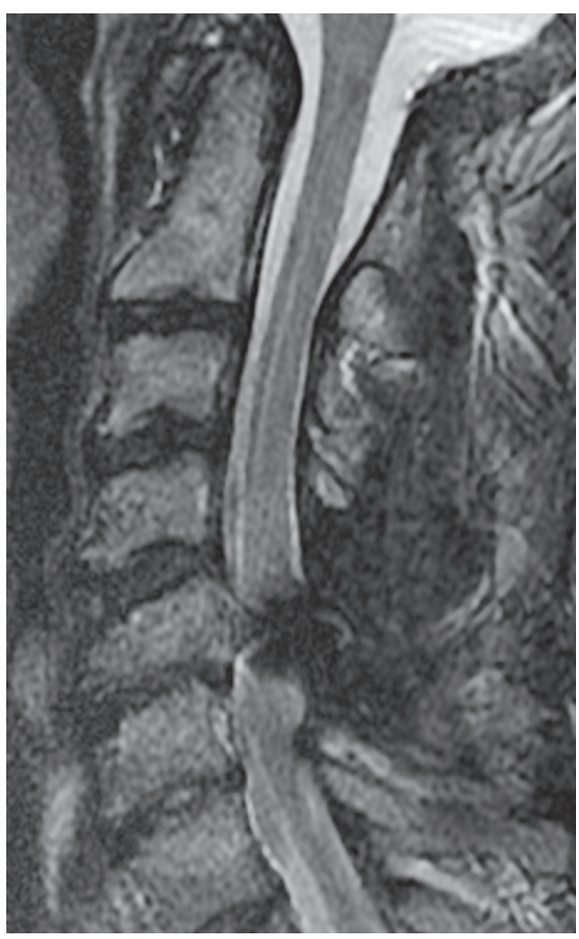

Figure 4 Sagittal T2 weighted MRI of the cervical spine performed 6 months postoperatively shows no persistent or recurrent dilated intrathecal flow voids and no T2 prolongation in the spinal cord (as limited at C5/6 by metallic artifact from surgical clips). A brain MRI performed at the same time (not shown) also demonstrated resolution of white matter lesions in the posterior fossa and corticospinal tracts.

lary veins intracranially: lateral medullary bridging veins connect the median posterior medullary vein to the lateral dural surface, draining either up toward the petrosal sinus or down toward the marginal sinus [1,8], just as the radiculomedullary veins connect the posterior midline spinal vein to the spinal epidural venous plexus. Given that the median posterior medullary vein ascends on the posterior medulla and divides just below the obex into the paired veins of the inferior cerebellar peduncle, it is also possible to describe the lateral bridging vein decompress-

ing the fistula in our patient as an inferior cerebellar peduncular bridging vein [9,10].

The degree of myelopathy due to SDAVF correlates with the number of spinal levels over which pressurized intrathecal veins course before exiting from the dura via bridging veins to the epidural venous plexus of the spine [4]. This case represents an extension of that phenomenon to the posterior fossa, with a pressurized midline posterior spinal vein continuing cranially as the medial posterior medullary vein that then decompresses via a posterior fossa bridging vein into a variant dural venous sinus above the foramen magnum that, in turn, flows out through the marginal sinus at the foramen magnum and subsequently into the condylar venous plexus. Initial angiographic misidentification of the fistula site as being in the posterior fossa (where the bridging vein entered the variant lateral linear dural venous sinus above the foramen magnum) led to surgical ligation of the bridging vein and resultant redirection of venous flow to anastomotic veins of the brainstem. As this is analogous to thrombosis, stenosis, or compression of venous outflow of a DAVF, phenomena thought to increase symptomatic venous hypertension in the spinal cord [4,11] and raise the risk of DAVF rupture intracranially [12], we deemed it appropriate to keep the patient's blood pressure low and promptly reoperate bilaterally at the C5/6 level to eliminate arterial inflow to the pressurized posterior midline spinal vein and its intracranial connections.

Conclusion

Bilateral cervical SDAVF with convergence on the posterior spinal vein and intracranial venous drainage can mimic a primary intracranial DAVF angiographically, particularly if high frame rate DSA is not performed. Knowledge of the anatomy of the cranial continuation of the posterior spinal vein as the median posterior medullary vein, and differentiation of bridging veins from the posterolateral spinal artery, may assist the angiographer in identifying the site of arteriovenous fistulization even in rare cases in which SDAVF are present bilaterally at the same spinal level.

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