

Cervical Spine Dural Arteriovenous Fistula With Coexisting Spinal Radiculopal Artery Aneurysm Presenting as Subarachnoid Hemorrhage: Case Report

Joshua W. Lucas, BS

Jesse Jones, MD

Azadeh Farin, MD

Paul Kim, MD

Steven L. Giannotta, MD

Department of Neurosurgery, Keck School of Medicine, University of Southern California, Los Angeles, California

Correspondence:

Joshua W. Lucas, BS,
1200 N State St,
Suite 3300,
Los Angeles, CA 90089.
E-mail: joshualu@usc.edu

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BACKGROUND AND IMPORTANCE: We present a patient with a cervical spine dural arteriovenous fistula associated with a radiculopal artery aneurysm at the same vertebral level presenting with subarachnoid hemorrhage.

CLINICAL PRESENTATION: A 45-year-old Native American man presented with sudden-onset severe headache, lethargy, and right hemiparesis. Computed tomography (CT) of the head showed subarachnoid hemorrhage and hydrocephalus. A subsequent CT of the neck showed an anterior spinal subdural hematoma from C2 to C4 causing mild cord compression. Carotid and vertebral angiography failed to demonstrate an intracranial aneurysm, but showed a spinal dural arteriovenous fistula originating from the right vertebral artery at the C5 neuroforamen. The severity of the patient's symptoms, atypical for rupture of a dural arteriovenous fistula, prompted more thorough angiographic evaluation. Thus, injection of the right thyrocervical trunk was performed, demonstrating a 4-mm spinal radiculopal artery aneurysm. Following ventriculostomy, a hemilaminectomy from C4 to C7 was performed with disconnection of the fistula from its drainage system. Subsequent resection of the aneurysm, which was determined to be the cause of the hemorrhage, was accomplished. The patient improved neurologically and was discharged to rehabilitation.

CONCLUSION: Spinal cord aneurysms from a separate vascular distribution may coexist with spinal dural arteriovenous fistulas. In the setting of spinal hemorrhage, especially in situations with an atypical clinical presentation, comprehensive imaging is indicated to rule out such lesions.

KEY WORDS: Aneurysm, Dural arteriovenous fistula, Subarachnoid hemorrhage

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Spinal dural arteriovenous fistulas (dAVFs) are the most common type of spinal vascular malformations, comprising 60 to 80% of such lesions.^{1,2} Spinal dAVFs are acquired lesions representing an abnormal connection between a dural branch of a radicular artery and vein along the spinal dural surface, forming in response to poorly understood pathophysiologic mechanisms.²⁻⁴ Most dAVFs are found at the neural foramina of the thoracolumbar junction, but they may arise at any spinal level.^{2,3}

ABBREVIATIONS: dAVF, dural arteriovenous fistulas; SAH, subarachnoid hemorrhage

Lesions in the thoracolumbar region manifest as progressive myelopathy with spastic paraparesis, sensory disturbances, or genitosphincteric dysfunction.^{1,3} Craniocervical spinal dAVFs can present with similar symptoms, but are distinguished by a propensity for causing subarachnoid hemorrhage (SAH) and cranial nerve dysfunction.^{3,5,6} Rupture of craniocervical dAVFs, although rare, typically produces low-grade subarachnoid hemorrhage. Here, we present an unusual case of a dAVF originating from branches of the vertebral artery and a coexisting aneurysm of the spinal pial circulation in a patient presenting with moderate-grade SAH.

CASE REPORT

A 45-year-old Native American man presented with sudden-onset severe headache. The patient also reported light sensitivity, neck pain and stiffness, nausea, and one episode of emesis. He was confused and lethargic with a Glasgow Coma Score of 13, and 2/5 strength in his right upper extremity with 4/5 strength in his right lower extremity. The patient admitted to a 10-year history of daily intravenous drug abuse. Of note, he denied a history of fevers, chills, or other symptoms of acute infection.

A noncontrast head computed tomography (CT) revealed extensive SAH in the upper cervical region and the basilar cisterns, and intraventricular hemorrhage with hydrocephalus (Figure 1). Subsequently, a CT angiogram of the head and neck to assess for a vascular etiology revealed an anterior spinal subdural hematoma extending from C2 to C4 causing mild cord compression (Figure 2). Also, a prominence of vessels surrounding the cord suggestive of a vascular malformation within the spinal canal was identified. A separate focus of hyperdensity at the C4/C5 level suggestive of an aneurysm was also noted (Figure 3). Magnetic resonance imaging of the cervical spine was strongly supportive of these findings (Figure 4).

Comprehensive cerebral and spinal cord angiography was subsequently performed. Injection of the right vertebral artery demonstrated an abnormally enlarged radicular artery at the C5 level and a dAVF at the C5 neuroforamen with a cranially directed draining vein extending to the suboccipital region (Figure 5). Based on the presence of posterior fossa SAH with an anterior spinal subdural hematoma, and the focus of hyperdensity seen on CT, injection of the right thyrocervical trunk was performed. This demonstrated a radiculopial artery aneurysm measuring 4 mm in diameter located anteriorly within the spinal canal at the level of the C4/C5 disc (Figure 6A and 6B). No intracranial vascular abnormalities or other anomalies were identified. These findings were consistent with a category IV subtype III spinal dAVF at the C5 neuroforamen and a radiculopial artery aneurysm at the level of the subdural hematoma fed by distal branches of the thyrocervical trunk.



FIGURE 1. Axial CT scan of the head showing subarachnoid hemorrhage and intraventricular hemorrhage.



FIGURE 2. Sagittal CT scan of the cervical spine with contrast demonstrating an anterior subdural mass extending from C2 to C4 and a focus of enhancement at the C4/C5 disc.

On postbleed day 4, the patient underwent a C4 to C7 right-sided hemilaminectomy with ligation of the dAVF and resection of the radiculopial artery aneurysm. Intraoperative angiography showed occlusion and absence of both lesions (Figure 7). The patient subsequently improved to a Glasgow Coma Score of 15 with 4+/5 strength in his right

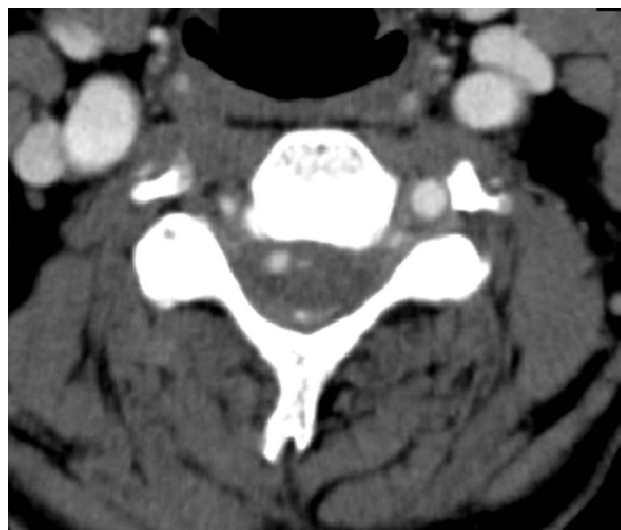


FIGURE 3. Axial CT scan with contrast through C4/C5 showing the anterior focus of enhancement later determined to be a radiculopial aneurysm and the posterior draining vein of the dAVF.



FIGURE 4. Sagittal T2-weighted magnetic resonance imaging of the cervical spine demonstrating a hyperintense ovoid lesion at the levels of C2 to C4 and multiple flow voids in the same location.



FIGURE 5. Injection of the right vertebral artery revealing a spinal dural arteriovenous fistula with a large feeding vessel originating from the level of C5 and a posterior cranially directed draining vein.

upper extremity and 5/5 strength in his right lower extremity, and was discharged to rehabilitation. Histologic evaluation of the aneurysm demonstrated an arterial wall without evidence of infection, excluding the possibility of a mycotic aneurysm or pseudoaneurysm.

DISCUSSION

Cranio-cervical dAVFs, although uncommon, have been well-characterized in the literature. SAH has been reported to occur in 34 to 45% of cases.^{2,5,6} When associated with hemorrhage, cases of dAVF rupture are usually characterized by a more benign clinical course than with aneurysm rupture. Fassett et al³ found that 95% of patients with cranio-cervical dAVF-related SAH presented with Hunt and Hess grade I or II hemorrhage, and no patients presented with Hunt and Hess grade IV or V hemorrhage. A large percentage of these patients had lesions with a cephalad or intracranially directed drainage pattern.^{5,6} Comparatively, 50 to 60% of patients with aneurysm-related SAH present with Hunt and Hess grade I or II hemorrhage, while 17 to 30% present with Hunt and Hess grade IV or V hemorrhage.^{7,8}

This patient, however, presented with moderate- to severe-grade SAH necessitating a ventriculostomy, clearly atypical for a cranio-cervical dAVF rupture. Upon discovery of the dAVF with injection of the right vertebral artery, this case could easily have been misdiagnosed as a dAVF alone. The discrepancy between

the patient's severity of SAH and that diagnosis, along with key findings on CT, heightened our suspicion that some other process might be responsible for the hemorrhage. Thus, injection of the right thyrocervical trunk was performed, demonstrating the coexisting radiculopial artery aneurysm.

We were confident, based on the anterior location of both the spinal subdural hematoma and the aneurysm, that the aneurysm was the source of the hemorrhage. The radiculopial artery aneurysm in this patient arose anteriorly from pial branches fed by the thyrocervical trunk. The dAVF, while located at the same vertebral level, arose posteriorly from right vertebral artery branches. Intraoperatively, the aneurysm was determined to be the lesion that ruptured based on its appearance and proximity to the anterior spinal subdural hematoma.

There is no definite explanation for the presence of the simultaneous lesions found in this patient. An association between dAVFs and aneurysms has only rarely been reported in the literature.⁹ However, other complex vascular structures such as arteriovenous malformations and multiple dAVFs have been found to coexist with dAVFs more frequently.¹⁰ As low-flow lesions, dAVFs rarely produce enough hemodynamic stress on the arterial walls to induce aneurysm formation of feeding arteries. Isolated aneurysms of the anterior spinal artery proper or

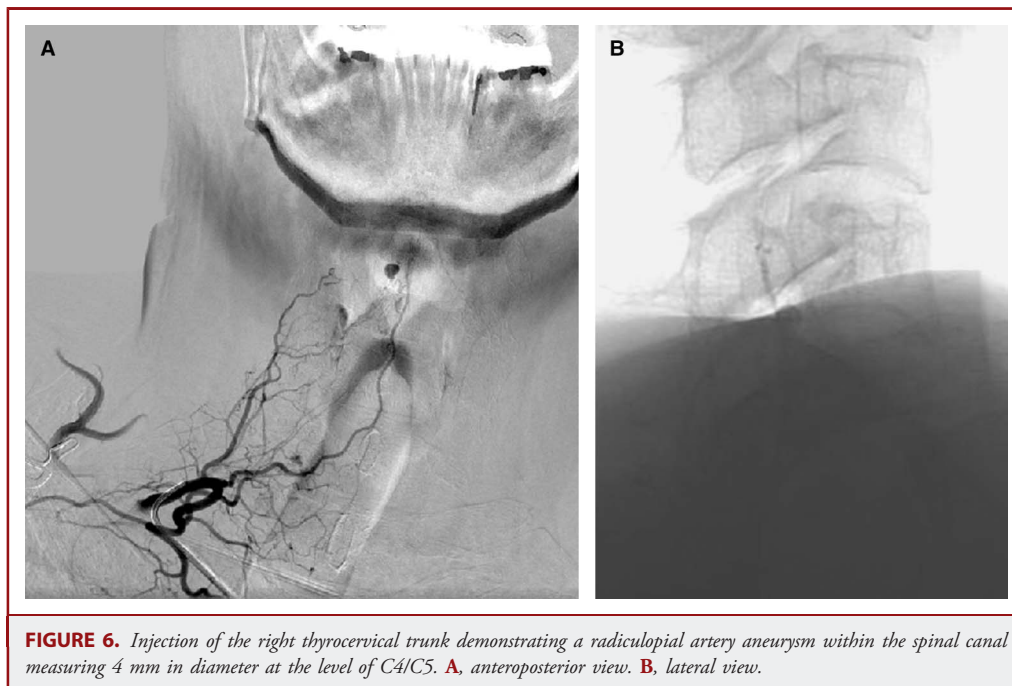


FIGURE 6. Injection of the right thyrocervical trunk demonstrating a radiculopial artery aneurysm within the spinal canal measuring 4 mm in diameter at the level of C4/C5. **A**, anteroposterior view. **B**, lateral view.

branches of the anterior spinal artery are also rare lesions, but when present are often associated with SAH.^{11,12}

The decision to proceed with an open microsurgical technique was made to allow for complete treatment of both the aneurysm and dAVF. The subdural hematoma was large enough to compress

the cervical spinal cord, so a longer laminectomy was performed to aid in decompression. Based on the patient's long history of intravenous drug abuse, we were suspicious that the radiculopial artery aneurysm might have actually been a mycotic aneurysm. However, no active infection was identified, and final pathological analysis of the aneurysm wall confirmed that the aneurysm was not mycotic.

The lesions in this patient highlight the possibility that craniocervical dAVFs and aneurysms of the spinal cord may coexist. Without additional clues from the CT scan and the patient's uncharacteristic presentation, the SAH and spinal hemorrhage might have mistakenly been attributed to rupture of the dAVF, leaving the true cause of the hemorrhage undiscovered. Thus, it is reasonable to conclude that early progression to more comprehensive vascular imaging studies in the diagnostic algorithm may be indicated to assess for the presence of such simultaneous lesions.

Disclosure

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

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FIGURE 7. Postoperative angiogram of the right vertebral artery displaying complete resolution of the spinal dural arteriovenous fistula.

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COMMENT

The authors present an interesting case of spinal dural arteriovenous fistula (SDAVF) presenting with intracranial subarachnoid (SAH) and cervical spinal subdural hemorrhage (SDH). In and of itself, SAH is a rare presenting symptom for SDAVF, although recently a clinical series from Toronto reported a relatively high incidence of hemorrhagic presentation of cervical DAVFs (Kim et al, *AJNR* 2010;31:1512-1515). Feeding artery aneurysms to a SDAVF have been reported previously by UCSF (Malek et al, *Neurosurgery* 1999;44:877-880), with pathologic correlation. The current case report also offers pathology confirming a true arterial aneurysm as opposed to a dissection or mycotic aneurysm.

This case underscores the importance of performing complete cervicocerebral angiography in order to delineate all possible feeding arteries to cervical spinal AV fistulas. Detailed angiography is essential prior to either endovascular or surgical treatment of these rare but important vascular malformations so that all arterial feeders to the AVF are identified as well as the pial supply to the spinal cord itself.

Steven W. Hetts
San Francisco, California