Transarterial Embolization of a Cervical Dural Arteriovenous Fistula Presenting with Subarachnoid Hemorrhage

S.A. ANSARI, J.P. LASSIG, E. NICOL, B.G. THOMPSON*, J.J. GEMMETE, D. GANDHI

Departments of Radiology and Neurosurgery, University of Michigan Health System, Ann Arbor, MI, U.S.A.*

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Summary

We describe a case of a 75-year-old man who presented with acute onset of headache and subarachnoid hemorrhage and initial cerebral angiography was deemed "negative". In retrospect, a faint contrast collection was present adjacent to the right vertebral artery at the C1 level suspicious for a small dural arteriovenous fistula (dAVF).

Follow-up angiography with selective microcatheter injections of the right vertebral artery and C1 radicular artery confirmed a complex dAVF with characteristically specific venous drainage patterns associated with a subarachnoid hemorrhage presentation. Subsequently, the cervical dAVF was treated with superselective glue embolization resulting in complete occlusion.

Cervical dAVFs are extremely rare vascular causes of subarachnoid hemorrhage. Both diagnostic angiography and endovascular treatment of these lesions can be challenging, especially in an emergent setting, requiring selective evaluation of bilateral vertebral arteries and careful attention to their cervical segments. Although only a single prior case of a cervical dAVF presenting with subarachnoid hemorrhage has been successfully treated with embolization, modern selective transarterial techniques may allow easier detection and treatment of subtle pathologic arteriovenous connections.

Introduction

Spinal dAVFs are the most common type (~80%) of spinal arteriovenous malformations, Anson-Spetzler Type 1, and predominantly involve the thoracolumbar spine in middle-aged males^{1,2}. Clinically, the patient's symptoms and signs are related to a slowly progressive myelopathy causing spastic or flaccid paraparesis, pain, sensory deficits and bladder/bowel sphincter dysfunction^{3,4}. In contrast, at the craniocervical junction and in the cervical spine, cervical dAVFs are rare lesions with approximately 50 reported cases in the literature³⁻¹⁰. A large subset of these cervical dAVFs (~40-45%) present with acute subarachnoid hemorrhage^{3,5-8}, rather than the dominant presentation of chronic myelopathy^{4,8,9}. More atypical presentations have included radiculopathy^{7,10} as well as distinctly unusual cases of tinnitus/cranial nerve VI palsy⁸, occipitalgia¹¹, transient ischemic attack¹², and a post-traumatic cervical dAVF leading to epidural hemorrhage and acute myelopathy¹³.

Case Report

A 75-year-old man with a history of prostate cancer and remote head trauma with loss of consciousness as a child presented to an outside emergency department with the "worst headache of his life", neck pain, and mild di-

Figure 1 Axial CT head image shows subarachnoid blood (arrowheads) predominantly at the foramen magnum and upper cervical spinal canal.

aphoresis. In addition, the patient described an intermittent episode of paresthesia in the right arm related to the sudden onset of severe suboccipital headache. The patient was neurologically intact, Hunt-Hess grade 2. The patient was transferred to our institution where CT head findings confirmed subarachnoid hemorrhage along the right lateral aspect of the foramen magnum and upper cervical spinal canal (figure 1). Cerebral angiography initially appeared negative, but during the procedure the right subclavian artery was injected using blood pressure cuff inflation instead of a selective right vertebral artery angiogram given its tortuous anatomy. Upon retrospective review, a small stagnant contrast collection was noted surrounding the right vertebral artery at the C1 level (figure 2). CT angiography of the head and neck and MR imaging of the total spinal axis were also unsuccessful in delineating the source of hemorrhage.

In order to evaluate the right vertebral artery definitively, a selective right vertebral artery angiogram via a right brachial artery approach was performed using a 5 French MPC Guide Catheter (Cordis Neurovascular, Miami,

FL) and revealed a small dAVF at the cranio-cervical junction. The fistula was supplied by the right C1 radicular artery branch, just superior to the C1 ring, with early opacification or drainage via the perivertebral venous plexus (figure 3). No additional arterial supply to the fistula from the external carotid, thyrocervical or costocervical circulation was identified. A superselective right C1 radicular arteriogram delineated the complex venous drainage of the dAVF with ascending intracranial venous drainage along the brainstem and a prominent cervical epidural venous plexus with associated venous varix or aneurysms (figure 4A and B). Due to the patient's age and small size of the fistula, the neurosurgical and interventional neuroradiology service opted for endovascular embolization. An Echelon-10 microcatheter (Micro Therapeutics, Irvine, CA) was advanced into the right C1 radicular artery and a 4:1 mixture of Ethiodol to N-butyl cyanoacrylate (NBCA) (Cordis Neurovascular, Miami, FL) was used to embolize the arterial feeder and proximal venous outflow of the dAVF. Post-embolization right vertebral artery angiogram demonstrated complete occlusion of the C1 dAVF (figure 5).

Following the procedure, the patient developed right sided numbness and mild weakness. MR imaging of the brain and cervical spine showed a punctuate focus of restricted diffusion in the right cerebellum. However, the patient's symptoms gradually resolved over two weeks with inpatient rehabilitation and outpatient physical therapy. In retrospect, this phenomenon was thought to be related to existing stenosis at the right vertebral artery origin and a thromboembolic complication during selective catheterization of the artery. A 12 month follow-up diagnostic cerebral angiogram showed no definite arteriovenous shunting or fistula.

Discussion

Spinal dAVFs classically arise from a single arterial feeder from the radicular arteries embedded in the dural sleeve of the proximal nerve root and drain through a radicular vein that communicates with the coronal venous plexus along the spinal cord, or less likely the epidural/paravertebral veins. The fistulous communication arterializes the perimedullary venous plexus and elevates the venous pressure

Figure 2 Lateral digital subtraction angiography (DSA) image of the right vertebral artery identifies suspicious area of subtle contrast puddling (arrow) adjacent to the right vertebral artery at the level of the C1 arch.

Figure 3 Anteroposterior DSA image of the distal right vertebral artery through a selective microcatheter injection demonstrates the C1 radicular branch (arrow) supplying the fistula as well as faint early opacification of the perivertebral venous plexus (arrowhead).

throughout the radial veins, decreasing the intramedullary arteriovenous gradient and therefore blood flow to the spinal cord^{3,5,14}. This venous congestion or hypertension eventually leads to patients experiencing symptoms of myelopathy or, in a subset of cervical dAVFs, acute subarachnoid bleeding^{3,5-8}. Some authors have proposed that insufficient venous drainage due to a lack of normal radicular veins predisposes to symptomatic congestion of the perimedullary venous plexus in patients with spinal dAVFs¹⁵. In cases of epidural venous drainage, extramedullary mass effect may further contribute to their myelopathic symptoms^{5,7,8,10}.

Aviv et Al recently performed an extensive review and analysis of the published literature on cervical dAVFs, highlighting several unique characteristics of these lesions. Cervical dAVFs maintain the age and sex demographics seen in thoracolumbar dAVFs, but the male:female ratio is lower 2-3:1 versus 7.5:1^{2,5}. The majority of these lesions (>80%) occur at the craniocervical junction and foramen magnum. Slightly less than half of patients (40-45%) with cervical

dAVFs present with acute subarachnoid hemorrhage instead of a progressive myelopathy, the predominant presentation in thoracolumbar dAVFs. In fact, thoracolumbar dAVFs have not been shown to present with subarachnoid hemorrhage in several large case series^{1-3,14,16-18}, unlike the hemorrhagic presentations of spinal intradural arteriovenous malformations or even pial/perimedullary arteriovenous fistulas. Although the exact etiology is unclear, it may be partly related to alternative venous pathways described in cervical dAVFs such as intracranial and/or epidural venous drainage^{3,5,7,8,10}. Hence, unique venous drainage patterns may be more susceptible to venous hypertension and bleeding.

Furthermore, specific and statistically significant venous drainage patterns were associated with subarachnoid hemorrhage. Almost all cervical dAVFs that presented with subarachnoid hemorrhage demonstrated either the presence of superiorly directed intracranial venous drainage or a venous varix^{3,5}. Consistent with these findings, our patient exhibited both intracranial venous drainage superiorly along the

Figure 4 Magnified anteroposterior (A) and lateral (B) DSA images of the C1 radicular branch through a superselective microcatheter injection delineates the exact location of the arteriovenous fistula (arrowheads), ascending intracranial venous drainage (black arrows), and prominent cervical epidural venous plexus with small associated venous varix or aneurysms (white arrows).

brainstem and a venous varix or aneurysms associated with the prominent epidural venous plexus (figure 4A and B). In contrast, Eskandar et al described five cervical dAVFs presenting with myelopathy in which venous drainage extended inferiorly without intracranial drainage in each case¹⁴.

Diagnostic angiography of dAVFs can be challenging at the craniocervical junction and in the cervical spine due to complex anatomy with very small and tortuous radicular arteries feeding the fistula. In an emergent subarachnoid hemorrhage setting, a cervical dAVF may not be identified due to spontaneous obliteration/thrombosis, mass effect/compression from the hemorrhage itself, or more likely because an inadequate study is performed while concentrating on the intracranial vascular anatomy. Interestingly, all of the reported cervical

dAVFs presenting with subarachnoid hemorrhage that were detected on initial cerebral angiography involved the craniocervical junction, which is usually included on standard views of the intracranial posterior circulation⁵. This demonstrates the value of including cervical views or at least carefully evaluating the craniocervical junction, the most common location of cervical dAVFs, during diagnostic angiography in the setting of an "occult" subarachnoid hemorrhage.

Conversely, six of the seven cases of cervical dAVFs presenting with subarachnoid hemorrhage that were negative on initial cerebral angiography arose from the right vertebral artery⁵. Moreover, all cervical dAVFs regardless of their presentation showed a tendency to be supplied by the right vertebral artery nearly twice as often as the left vertebral artery⁵. In

many practices, selective right vertebral artery injection is avoided in cerebral angiography if sufficient reflux from the commonly dominant left vertebral artery can opacify the right posterior inferior cerebellar artery. Our case exemplifies the importance of a selective right vertebral artery injection as the C1 dAVF was nearly missed on initial angiography and perhaps only detected due to its craniocervical location. Therefore, in a patient presenting with a subarachnoid hemorrhage from an indeterminate cause, a diligent search for a cervical dAVF would be prudent. It is imperative to perform selective four-vessel angiography, the bilateral carotid and vertebral arteries, with attention to the craniocervical junction. If this initial work-up is negative, cervical views and further selective injections of the external carotid arteries, thyrocervical and costocervical trunks should be considered.

Both microsurgical and endovascular treatment of spinal dAVFs depend on disrupting the AVF from its spinal venous drainage, thereby decreasing the venous plexus pressure and restoring normal perfusion to the spinal cord. The optimal treatment strategy is controversial and a decision should probably be deferred to the unique parameters or anatomy of each case. Spinal dAVFs with complex and tortuous arterial feeders may not be amenable to super-selective catheterization or distal passage of embolic material. Although embolization may still be possible in spinal dAVFs supplied by radicular arteries in proximity to the origins of radiculomedullary/radiculopial arteries, such as the artery of cervical enlargement or artery radicularis magna (artery of Adamkiewicz), the risks and concerns for iatrogenic injury or embolic reflux into crucial vasculature supplying the anterior/posterior spinal arteries must be balanced with the anatomy and clinical context of each patient.

Approximately 40% of patients will eventually require surgery due to inadequate embolization or fistula recurrence, but definitive endovascular treatment of spinal dAVFs has been shown in several studies^{14,16,17}. Successful embolization of spinal dAVFs requires embolic material such as liquid acrylic or "glue" (NBCA) to penetrate into the arteriovenous connection and proximal draining vein, preventing recanalization or further recruitment by the fistula and hence recurrence.

Following embolization of the spinal dAVF,

Figure 5 Magnified lateral DSA image of the right vertebral artery after embolization demonstrates complete occlusion of the fistula and no evidence of early opacification of the perivertebral epidural venous plexus or ascending perimedullary venous drainage.

angiography should confirm the obliteration of the fistula while preserving normal spinal venous drainage¹⁴.

Embolization of cervical dAVFs has also been described by multiple groups^{10,11,13,14,19}, but only one case of a cervical dAVF presenting with subarachnoid hemorrhage has successfully been treated⁷. Cahan et Al had embolized this C5 dAVF using a single detachable balloon, but this strategy has largely been replaced by glue embolization to prevent fistula recurrence as mentioned above. Willinsky et Al attempted embolization with NBCA of a C8 dAVF after rapid development of quadriplegia and bladder/bowel dysfunction following an "occult" episode of subarachnoid hemorrhage seven months earlier⁸. Unfortunately, recurrence of the fistula required surgical clipping of the intradural vein two months later. Recently, Vankan et Al demonstrated coil embolization of a post-traumatic C1 dAVF after a C1-C2 fracture, but it presented eight months afterwards with a rapidly progressive myelopathy due to a C1-C5 epidural hemorrhage rather than subarachnoid hemorrhage¹³.

Conclusions

We have described a unique case of a cervical dAVF presenting with subarachnoid hemorrhage and its successful treatment with transarterial embolization. It highlights the need for selective angiography of bilateral vertebral arteries, especially in a setting of an "occult" subarachnoid hemorrhage. In addition, since the majority of cervical dAVFs occur at the cranio-

cervical junction and can be very difficult to diagnose, careful attention to this region is warranted to optimize their detection. Once these lesions are recognized, specific venous drainage patterns may help predict their clinical presentation and/or direct appropriate treatment. With rapid improvements in catheter technology and embolic materials, superselective access to dAVFs and their endovascular disconnection will more likely be possible.

References

- Kendall BE, Logue V: Spinal epidural angiomatous malformations draining into intrathecal veins. *Neuroradiology* 13(4): 181-189, 1977.
- Symon L, Kuyama H, Kendall B: Dural arteriovenous malformations of the spine. Clinical features and surgical results in 55 cases. *J Neurosurg* 60(2): 238-247, 1984.
- Kinouchi H, Mizoi K et Al: Dural arteriovenous shunts at the craniocervical junction. *J Neurosurg* 89(5): 755-761, 1998.
- Reinges MH, Thron A et Al: Dural arteriovenous fistulae at the foramen magnum. *J Neurol* 248(3): 197-203, 2001.
- Aviv RI, Shad A et Al: Cervical dural arteriovenous fistulae manifesting as subarachnoid hemorrhage: report of two cases and literature review. *Am J Neuroradiol* 25(5): 854-858, 2004.
- Do HM, Jensen ME et Al: Dural arteriovenous fistula of the cervical spine presenting with subarachnoid hemorrhage. *Am J Neuroradiol* 20(2): 348-350, 1999.
- Cahan LD, Higashida RT et Al: Variants of radiculomeningeal vascular malformations of the spine. *J Neurosurg* 66(3): 333-337, 1987.
- Willinsky R, TerBrugge K et Al: The variable presentations of craniocervical and cervical dural arteriovenous malformations. *Surg Neurol* 34(2): 118-123, 1990.
- Trop I, Roy D et Al: Craniocervical dural fistula associated with cervical myelopathy: angiographic demonstration of normal venous drainage of the thoracolumbar cord does not rule out diagnosis. *Am J Neuroradiol* 19(3): 583-586, 1998.
- Kohn M, Takahashi H et Al: A cervical dural arteriovenous fistula in a patient presenting with radiculopathy. Case report. *J Neurosurg* 84(1): 119-123, 1996.
- Chiba S, Nishioka H et Al: Cervical dural arteriovenous malformation presenting with right-sided occipitalgia: before and after successful treatment by embolization. *Headache* 34(4): 234-236, 1994.
- Masuo O, Ozaki F et Al: Dural arteriovenous fistula at the cranio-cervical junction presenting transient ischemic attack: a case report. *No Shinkei Geka* 27(11): 1043-1046, 1999.
- Vankan Y, Demaerel P et Al: Dural arteriovenous fistula as a late complication of upper cervical spine fracture. Case report. *J Neurosurg* 100: 382-384, 2004.
- Eskandar EN, Borges LF et Al: Spinal dural arteriovenous fistulas: experience with endovascular and surgical therapy. *J Neurosurg* 96: 162-167, 2002.
- Hassler W, Thron A, Grote EH: Hemodynamics of spinal dural arteriovenous fistulas. An intraoperative study. *J Neurosurg* 70(3): 360-370, 1989.
- Mourier KL, Gelbert F et Al: Spinal dural arteriovenous malformations with perimedullary drainage. Indications and results of surgery in 30 cases. *Acta Neurochir (Wien)* 100(3-4): 136-141, 1989.
- Niimi Y, Berenstein A et Al: Embolization of spinal dural arteriovenous fistulae: results and follow-up. *Neurosurgery* 40(4): 675-682, 1997.
- Berenstein A, Lasjaunias P: *Surgical Neuroangiography 5: Endovascular Treatment of Spine and Spinal Cord Lesions*. Berlin: Springer 5-24, 1992.
- Mascalchi M, Scazzari F et Al: Dural arteriovenous fistula at the craniocervical junction with perimedullary venous drainage. *Am J Neuroradiol* 17(6): 1137-1141, 1996.

Sameer Ansari, M.D., Ph.D.
Division of Interventional Neuroradiology
Department of Radiology
University of Michigan Health System
1500 East Medical Center Drive, B1D330A
Ann Arbor, MI 48109-0030, U.S.A.
E-mail: sameansa@med.umich.edu