Intracranial subarachnoid hemorrhage resulting from cervical spine dural arteriovenous fistulas: literature review and case presentation

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Cervical dural arteriovenous fistulas (dAVFs) are a rare cause of intracranial subarachnoid hemorrhage (SAH) but should be considered when other sources are not found. Subarachnoid hemorrhage caused by dAVF is thought to occur as a result of venous hypertension in most cases. The clinical presentation, acute onset of severe headache, is similar to that in patients with other causes of SAH; however, severe neurological deficits (Hunt and Hess Grade IV and V SAH) have not been reported in SAH caused by cervical dAVFs. Patients with this type of SAH commonly report suboccipital headache, neck pain, and nausea, and thus these hemorrhages can be easily dismissed as perimesencephalic SAH. Vigilant evaluation with 4-vessel cerebral angiography, including selective catheterization of both proximal vertebral arteries, should be performed. The practice of unilateral vertebral artery injection with reflux into the contralateral vertebral and posterior inferior cerebellar arteries has the potential to overlook cervical dAVF. Magnetic resonance imaging may be useful to evaluate for other causes of SAH but is probably not sensitive for the identification of a cervical dAVF. Surgical treatment of this lesion has an excellent outcome.

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Key Words • cervical spine • dural arteriovenous fistula • intracranial subarachnoid hemorrhage

Spinal dAVFs are acquired vascular lesions characterized by an abnormal connection between a dural branch of a radicular artery and a radicular vein along the spinal dural surface. The fistulous connection is most commonly identified near the intervertebral foramen along the dorsal aspect of the nerve root near the axilla of the dural sleeve. Spinal dAVFs may arise at any level from the foramen magnum to the sacrum, but are most commonly found in the thoracolumbar junction. Patients characteristically present with progressive myelopathy, including spastic paraparesis and bladder dysfunction resulting from venous hypertension in the medullary veins and the pial coronary venous plexus of the spinal cord.1,4,11,17,19

Cervical dAVFs represent only a minority of spinal dAVFs, but in contrast to their thoracolumbar counterparts, they may be associated with protean clinical manifestations such as SAH, vascular congestive myelopathy, radiculopathy, and cranial nerve dysfunction.2,12 In 1987, Cahan et al.3 published the first report of an SAH occurring secondary to cervical dAVFs, and a number of small case series have been published subsequently. Aviv and colleagues2 reported that 45% of cervical dAVFs in the literature were in patients who presented with SAH. In this review, we focus on SAH associated with cervical dAVFs, including the clinical presentation, radiographic evaluation, and treatment.

Methods

The data from our single patient combined with a PubMed review of the English literature yielded 22 cases of SAH associated with cervical spinal dAVF (Table 1).2,3,5,7,12,14,16,18,21 Data regarding symptoms at presentation, location of dAVF, treatment, and outcome were collected.

Results

Clinical Presentation

In patients for whom the clinical history was reported (18 of 22 patients), the majority presented with an H &
Grade II hemorrhage and either headache, neck pain, or nausea (Table 1). Only 1 of 18 patients had neurological deficits (mild right hemiparesis) noted on initial evaluation. One other patient, reported on by Liu et al., presented with headache only, but rapidly progressed to quadriplegia within a few hours of presentation because of presumed spinal cord compression from a hematoma. No patients presented with H & H Grade IV or V hemorrhages.

### Diagnostic Evaluation

All patients were noted to have an intracranial SAH, and in most instances, this was noted on CT scanning. Angiography was performed in 21 of 22 patients. In a case described by Niwa et al., angiography was not performed, but a dAVF was noted on intraoperative exploration.

In 5 patients, angiography failed to reveal the fistula on initial evaluation, and in at least 3 of these 5, the chosen angiography technique contributed to the failure of diagnosis. In these 3 cases (Do et al., Aviv et al., and the present study), angiography with selective catheterization of a single VA was performed. The contralateral distal VA and PICA were evaluated with a reflux of contrast material into these vessels. The use of this technique meant that the proximal VA was not evaluated and therefore that the dAVF arising from it was not identified.

In the remaining 2 cases in which initial angiography failed to reveal a fistula, detailed angiography findings were not provided. Willinsky et al. reported a 7-month delay in diagnosis in a patient who presented with an SAH. Progressive myelopathy, presumably from venous congestive myelopathy related to the dAVF, developed in this patient after the initial evaluation failed to find the cervical dAVF. Kinouchi et al. described a patient with a recurrent SAH and cervical dAVF who had initially presented with an angiographically negative SAH 10 years earlier.

### Treatment and Outcome

Endovascular techniques have been used in only 2 reported cases and in 1 of these treatment was incomplete and surgery was required for definitive ligation. One patient died of complications associated with a CSF shunt placement procedure before dAVF treatment could be performed. In 20 patients, the dAVF was treated with surgical ligation of feeding artery or draining vein. In 19 of these cases, surgery was the primary treatment, and in 1 case, surgery was performed after failure of endovascular treatment. Angiographic follow-up with complete obliteration of the dAVF was noted in many cases, and no case of recurrence after surgical closure has been reported.

Patient outcome was reported in 20 of 22 cases. In

### Table 1: Summary of reported cases of intracranial SAH cases resulting from cervical dAVFs

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Location</th>
<th>Presenting Symptoms</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cahan et al., 1987</td>
<td>C-5</td>
<td>NA</td>
<td>embo</td>
<td>NA</td>
</tr>
<tr>
<td>Willinsky et al., 1990</td>
<td>C-8</td>
<td>mild right arm &amp; leg weakness</td>
<td>embo (delayed); later surgery</td>
<td>2</td>
</tr>
<tr>
<td>Morimoto et al., 1992</td>
<td>C-5</td>
<td>headache &amp; dysesthesias in arm</td>
<td>surgery</td>
<td>1</td>
</tr>
<tr>
<td>Niwa et al., 1997</td>
<td>Oc–C1</td>
<td>headache</td>
<td>surgery</td>
<td>1</td>
</tr>
<tr>
<td>Kinouchi et al., 1998</td>
<td>C-1</td>
<td>headache</td>
<td>surgery</td>
<td>1</td>
</tr>
<tr>
<td>Do et al., 1999</td>
<td>C-1</td>
<td>headache &amp; nausea</td>
<td>surgery</td>
<td>1</td>
</tr>
<tr>
<td>Hashimoto et al., 2000</td>
<td>C-1</td>
<td>headache &amp; nausea</td>
<td>surgery</td>
<td>1</td>
</tr>
<tr>
<td>Hida et al., 2002</td>
<td>C2–3</td>
<td>NA</td>
<td>surgery</td>
<td>improved</td>
</tr>
<tr>
<td>C-3</td>
<td>NA</td>
<td>surgery</td>
<td>improved</td>
<td></td>
</tr>
<tr>
<td>C-5</td>
<td>headache</td>
<td>surgery</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>C-6</td>
<td>NA</td>
<td>surgery</td>
<td>unchanged</td>
<td></td>
</tr>
<tr>
<td>Aviv et al., 2004</td>
<td>C-1</td>
<td>headache &amp; confusion</td>
<td>died before treatment</td>
<td>died</td>
</tr>
<tr>
<td>C-2</td>
<td>headache &amp; vomiting</td>
<td>surgery</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Liu et al., 2008</td>
<td>C-5</td>
<td>headache/neck pain (progressed to quadriplegia w/in hrs)</td>
<td>surgery</td>
<td>NA</td>
</tr>
<tr>
<td>Fassett et al., 2008</td>
<td>C-2</td>
<td>headache &amp; neck pain</td>
<td>surgery</td>
<td>1</td>
</tr>
</tbody>
</table>

* Numbers represent Glasgow Outcome Scale scores. Abbreviations: embo = embolization; NA = not available; Oc = occiput.
15 of 16 patients for whom details regarding outcome were provided, there was an excellent outcome without apparent residual deficits or disabilities. One patient was noted to have some mild residual hemiparesis after treatment. Two patients were reported as improved, and 1 as unchanged, but it was unclear whether these 3 had any residual disabilities at follow-up. Vasospasm was not reported as a complication or finding in any of the reported cases.

**Case Report**

This 78-year-old right-handed man presented with the sudden onset of acute occipital headache that arose while he was taking a bath. His medical history was significant for bifrontal decompressive craniectomy for severe traumatic brain injury when he was 70 years of age. His neurological examination was no different from his baseline state, with an established right visual field deficit attributable to posttraumatic gliosis of the left occipital lobe, and mild dysphasia. Computed tomography scanning of his head revealed a hemorrhage in a similar pattern to that characteristic of a benign perimesencephalic SAH. Subarachnoid hemorrhage was noted in the prepontine cistern, bilateral ambient cisterns, and left proximal sylvian fissure (Fig. 1). A CT angiogram of the head did not reveal any vascular abnormalities, but this study did not include the cervical spine. A diagnostic cerebral angiogram, consisting of bilateral carotid artery and left VA injections, failed to reveal any underlying vascular pathological conditions either. The right VA was not selectively catheterized because of distal retrograde filling and opacification of the right PICA during injection of the left VA. The patient had a stable clinical course in the intensive care unit for the next 7 days with no clinical evidence vasospasm. Repeated angiography was performed on Day 7. Selective catheterization of the right VA this time showed slow arteriovenous shunting from the radiculomeningeal branch of the C-2 radicular artery to a radicular vein, which was associated with a prominent variceal enlargement, and finally to the epidural venous plexus (Fig. 1B). The patient underwent a far lateral suboccipital craniectomy. The
right VA was dissected free from the level of the atlas to the axis, and a unilateral C-1 and C-2 laminectomy was performed. The radiculomeningeal branch of the C-2 radicular artery was ligated flush with the dura mater at the level of the C-2 nerve root sleeve. Abnormal, blister-like vasculature was noted to protrude from the dura at the level of the nerve root sleeve and was successfully obliterated with bipolar cauterization. Intradural exploration did not reveal any further abnormal vasculature. The patient tolerated the procedure well and no further neurological deficits developed; he was discharged home on postoperative Day 5. Postoperative angiography confirmed successful elimination of the fistula (Fig. 1D).

Discussion

Subarachnoid hemorrhage is almost never associated with a dAVF of the thoracic and lumbosacral spine.23 Cervical dAVFs, however, although uncommon, are associated with a high incidence of hemorrhagic presentation. In their review, Aviv et al.2 found SAH to be a presenting event in 45% of the cervical dAVF cases reported in the literature.

The mechanism underlying hemorrhagic presentation is presumed to be attributable to venous hypertension, but is not entirely understood. Arteriovenous shunting results in arterialized blood entering the medullary veins and the valveless pial coronal venous plexus and radial veins. The apparent higher incidence of hemorrhage in cervical dAVFs compared with thoracolumbar dAVF is difficult to explain. Aviv and colleagues2 noted that variceal enlargement of the medullary veins was significantly more common in patients with cervical dAVF and SAH than in patients with a nonhemorrhagic presentation. Moreover, the authors concluded that cephalad or intracranially directed venous drainage was significantly associated with SAH. Cephalad drainage pathways are considered abnormal, presumably resulting from thrombosis of normal drainage pathways, which are usually caudally directed, above the level of the heart.2

Although SAH resulting from a cervical dAVF is rare, it should be considered in the differential diagnosis, especially when the initial angiographic evaluation is negative for spontaneous SAH. In our patient (Fig. 1), reliance on retrograde filling of the right VA and PICA from a left VA injection led to failure of opacification of the fistula. Not uncommonly, cervical dAVFs do have feeding arteries from the right VA. The right VA was found to supply a cervical dAVF in 63% of patients with SAH and 58% of patients without SAH in the review of Aviv and associates.2 Our case report underscores the importance of meticulous angiographic investigation in cases of suspected cervical dAVF. Evaluation should include selective injections of both external carotid and vertebral arteries, supplemented by dedicated views of the cervical vertebral arteries. Some centers also have protocols to obtain an MR imaging of the cervical spine in all cases of angiographically negative SAH, which has the potential of identifying vascular anomalies such as dAVF.

Subarachnoid hemorrhage caused by a cervical dAVF has a more benign clinical presentation and than aneurysmal SAH. In 20–30% of cases, patients with aneurysmal SAH present with H & H Grade IV and V hemorrhage.3,10,12 In contrast, 95% of the patients included in our combined series of cervical dAVF-related SAH present with H & H Grades I and II hemorrhage. There have been no reported cases of Grade IV or V SAH occurring as a result of cervical dAVF. The clinical course after the initial hemorrhage also appears to be more benign compared with aneurysmal SAH, with no cases of vasospasm reported in these patients. There was only 1 death among the cases reviewed, and it was the result of CSF “shunt complications.” The more benign clinical course resembles that of paramesencephalic SAH.20

Treatment of dAVFs presenting with SAH has predominantly been surgical. Only 2 published cases have been treated endovascularly, and in 1 of these cases, surgical ligation was ultimately required because of progressive symptoms and a residual dAVF. Surgery appears to be very effective with ligation of the feeding artery or draining vein. No cases of recurrence have been reported after surgical intervention. If left untreated, it appears that these fistulas have the potential to cause recurrent SAH as demonstrated by the case reported by Kinouchi et al.,12 and there is also a risk of congestive venous myelopathy.

Conclusions

Cervical dAVFs can cause intracranial SAH and may be overlooked without careful angiographic evaluation that includes both proximal VAs. Subarachnoid hemorrhage resulting from a cervical spine dAVF has a benign clinical course and an overall good prognosis when effectively treated.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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References


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