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Case Report

Cervical Dural Arteriovenous Fistulae Manifesting as Subarachnoid Hemorrhage: Report of Two Cases and Literature Review

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Summary: Dural arteriovenous fistulas (DAVFs) in the craniocervical junction are rare but clinically important. DAVFs can be associated with subarachnoid hemorrhage (SAH), a feature distinguishing them from DAVFs in the thoracolumbar region. These lesions are often overlooked at cerebral angiography performed to assess SAH and account for a small proportion of angiographically negative SAHs. After managing two cases of cervical spinal DAVF manifesting as SAH, we analyzed all cases in the literature to identify features associated with bleeding at presentation.

Spinal dural arteriovenous fistulas (DAVFs) are small arteriovenous fistulas situated within the dura layers. They most commonly occur at the dorsal thoracolumbar junction in middle-aged men (1–3) and rarely bleed (1). Presenting symptoms and signs are progressive myelopathy with spastic paraparesis, sensory disturbances, and bowel and bladder dysfunction (4). In a number of patients, DAVFs are situated in the cervical spine and at the craniocervical junction. These lesions differ from their counterparts in the thoracolumbar region and have a wider mode of presentation, including subarachnoid hemorrhage (SAH) (4–14), radiculopathy (5, 15), myelopathy (2, 4, 5, 11, 13, 14, 16, 17–24), and cranial-nerve disturbance (14). We report two additional cases of cervical spinal DAVFs (cDAVFs) manifesting with SAH. We discuss morphologic features that appear to be associated with a presentation of SAH and review the current literature.

Case Reports

Case 1

A 57-year-old man was admitted to the hospital after a sudden onset of severe headache and confusion. On admission, his Glasgow Coma Scale score was 14, and physical examination revealed no focal neurologic deficit. CT of the head showed hydrocephalus, as well as hemorrhage in the basal cisterns and in the third and fourth ventricles. The hydroceph-

alus was managed by external ventricular drainage. Cerebral angiography was performed the next day with injection of both internal carotid arteries and the right vertebral artery. Good reflux into the distal left vertebral artery filled the posterior inferior cerebellar artery. The angiograms demonstrated no evidence of arteriovenous malformation or intracranial aneurysm. Subsequent T2-weighted MR imaging showed increased signal intensity in the medulla and upper cervical cord and also a small hematoma posterolateral to the cord at the level of the atlas. Repeat angiography performed 2 weeks later showed a spinal cDAVF at the C1 level just below the foramen magnum. This was fed by the C1 dural branch of the left vertebral artery and drained via an enlarged epidural vein into the extravertebral veins. A large varix was present behind the spinal cord on the left at the C1 level. Surgical treatment was planned, but shunt-related complications developed and the patient died.

Case 2

A 53-year-old woman presented after experiencing a sudden occipital headache with vomiting and then collapsing. Her Glasgow Coma Scale score was 15, with no focal neurologic deficit upon her arrival at the hospital. Head CT showed SAH in the posterior fossa, with blood in the cisterna magna and fourth ventricle. Four-vessel cerebral angiography showed irregularity in the distal right vertebral artery. MR imaging revealed an intradural extramedullary hematoma anterolateral to the cord behind the C2 vertebral body. Repeat angiography performed a week later revealed an irregular right vertebral artery and a cDAVF supplied by the right C2 dural branch of the vertebral artery, with a large venous varix. Drainage was via an enlarged epidural venous plexus. Three weeks after admission, the patient underwent upper cervical laminectomy at C1-C3, with coagulation and division of the cDAVF. After the dura was opened, large arterial feeding vessels were seen running into the varix, which lay under the right C2 nerve root. These vessels were carefully dissected off the nerve root, coagulated, and divided. After surgery, the patient developed persistent left-hindquarter numbness but otherwise made a good recovery. She was discharged from hospital on her 8th postoperative day. Follow-up MR imaging performed at 6 months showed no evidence of residual cDAVF.

Literature Review

We performed a literature review of all peer-reviewed journals, including foreign-language journals, by using PubMed and Medline. The following keywords were used: *cervical, dural,* and *arteriovenous fistula*. We found 42 cases and extracted the following data from them: presence of SAH, initial angiographic result, vascular supply and drainage, presence of varices and/or aneurysms, and direction of drainage. The data were analyzed by using SPSS for Win-

Key Features of Reviewed Literature

Author and Year	SAH	Initial Angiographic SAH	Feeder Vessels	Venous Drainage	Varix
Symon et al, 1984 (2)	No	NA	Meningeal br VA	CVP	No
Cahan et al, 1987 (5)	No	NA	R-VA, ACA, DCA	Extradural	No
	No	No	C5 R-VA	CVP, extradural	No
Oda et al, 1989 (16)	No	NA	R-VA	CVP, MV	No
Willinsky et al, 1990 (14)	No	NA	Neuromeningeal br APA	CVP, intracranial	No
	No	CCA, R-VA	CVP	No	
		NA	TCA, CCA	Extradural	Yes
		NA	Neuromeningeal br APA, OCA	Cavernous sinus, cortical vein over temporal lobe	No
Gaensler et al, 1990 (17)	No	NA	R-VA, neuromeningeal br of APA	CVP	No
		NA	R-VA, neuromeningeal br of APA, PICA	CVP	No
Revierz et al, 1991 (13)	No	NA	R-VR, OCA	CVP, spinal veins	No
		Yes	R-VA	CVP, RMV	No
Yamada et al, 1991 (18)	No	NA	R-VA, TCA, CCA	Extradural	No
Morimoto et al, 1992 (10)	Yes	No	C5 R-VA	MV, CVP, rostral	No
Glasser et al, 1993 (19)	No*	NA	R-VA	CVP	Yes
Ikeda et al, 1994 (9)	Yes	No	R-VA	Extradural, MV (Int vert ven pl)	Yes
Kohno et al, 1996 (15)	No	NA	R-VA, TCA, CCA	Internal jugular veins	No
Mascalashi et al, 1996 (20)	No	NA	APA, M-VA	RMV	No
		NA	APA	RMV	No
Niwa et al, 1997 (11)	Yes	Yes	C1 RA, postmeningeal	CVP, RMV	No
		NA	C1 RA, postmeningeal	CVP	No
Trop et al, 1998 (25)	No	NA	M-VA	CVP, MV	No
Hiroyuki et al, 1998 (26)	Yes	Yes?	C1 RA	Intracranial, CVP	Yes
		Yes?	C1 RA	Intracranial	Yes
		NA	C1 RA, OCA	CVP	No
		NA	C1 RC	Intracranial, CVP	No
		No	C1 and C2 RAs	Intracranial	Yes
		Yes?	C1 RA	Intracranial	Yes
		Yes?	C1 RA	Intracranial	No
		Yes?	C1 and C2 RAs	Intracranial	No
		Yes?	C1 RA	Intracranial, CVP	No
		NA	C2 RA	Intracranial	No
Do et al, 1999 (6)	Yes	No	Muscular br VA	MV, rostral	No
Hanabusa et al, 1999 (7)	Yes	Yes?	Meningeal branch of VA	Intracranial	No
		Yes?	Meningeal branch of VA	Intracranial	No
Yoshida et al, 1999 (21)	No	NA	Direct fistula between VA/spinal veins at occipital C1	CVP, MV	No
Oishi et al, 1999 (22)	No	NA	VA	Spinal veins	No
Miyoshi et al, 1999 (23)	No	NA	VA	Spinal veins	No
Nonaka et al, 1999 (12)	Yes	No	C1–C2 R-VA	Extradural, CVP	No
Hashimoto et al, 2000 (8)	Yes	Yes?	R-VA	Intracranial	No
Reinges 2001 (24)	No	Yes	VA	MV	No
	No	Yes	VA	MV	No
	No	Yes	VA	MV	No
Aviv et al, 2001, current report	Yes	No	R-VA	Extradural	Yes
		Yes	C1–C2 R-VA	Extradural	

dows, version 9.0 (SPSS Inc, Chicago, IL). The Fisher exact test was used to compare proportions between the SAH and non-SAH groups, and the Wilcoxon rank-sum test was used to compare distributions of continuous variables. Statistical significance was defined as $P < .05$.

The Table summarizes the cases of cDAVFs in the literature, listing the presence of SAH, initial angiographic result, vascular supply and drainage, and the presence of varices.

Forty-four cases of cDAVF were described as of this writing. Symon et al published the first report in 1984 (2), and Kinouchi et al reported the largest series, comprising 10 patients, in 1998 (4). Twenty patients (45%) of 44 presented with SAH. The remaining patients presented with myelopathy (19 [79%] of 24), radiculopathy (two [8%] of 24), and tinnitus and palsy of cranial nerve VI (one [4%] of 24) (14). Two incidental discoveries (8%) were made after imaging for alternate pathologic condition (4). In the 19 patients presenting with myelopathy, bladder and/or bowel dysfunction was present in 12 (63%) (2,

4, 14, 16, 17, 20, 23, 24). Three patients were excluded from further analysis: One had a cerebellar arteriovenous malformation in addition to a dural fistula at C1, another had a sigmoid sinus dural fistula in addition to a dural fistula at C1, and the final patient had an epidural venous fistula at C4-C5. Therefore, our remaining analyses pertained to 41 subjects.

The median age of presentation of patients with cDAVF was 58 years (range, 19–74 years). This group included 29 men (71%) and 12 women (29%). The median age of presentation for the 20 patients with SAH was 60.5 years (range, 19–70 years); this was similar to the age of the 21 patients in the non-SAH group (median, 57 years; range, 44–74 years; P = 1.0). The cDAVF was at the craniocervical junction in 20 patients (49%), the foramen magnum in 15 (37%), between C3 and C6 in five (12%), and at C8 in one (2%). There was no significant difference in patient sex (P = .99) or the cervical level of the cDAVF (P = 1.0) between the SAH group and the non-SAH group.

The side of the vertebral radicular-branch arterial supply was reported is some cases. In the SAH group, it was from the right in 12 (63%) of 19 patients, and in the non-SAH group, it was from the right in 11 (58%) (P = .99). The right vertebral artery supplied the cDAVF almost twice as often as the left vertebral artery in both groups.

In the SAH group, superiorly directed drainage occurred in 12 (60%) of 20 patients (4, 7, 8). In 10, the drainage was intracranial, but in two, it did not reach the cranium (6, 10). The non-SAH group had two DAVFs (14) with intracranial drainage (10%). Intracranial or superior drainage is significantly associated with a presentation with SAH (P = .002).

The presence of a varix was significantly associated with a presentation with SAH (P = .02) (14). A venous varix or pouch was present in seven (35%) patients with SAH (4) and in one (5%) patient in the non-SAH group. The pathway for venous drainage in the SAH group was via the coronal venous plexus in eight (40%) patients, epidural in five (25%), and intracranial in 10 (50%). Drainage in the non-SAH group was via the coronal venous plexus in 18 (86%), epidural in four (19%), and intracranial in one (5%). Apart from intracranial drainage, the pattern of venous drainage was not associated with a presentation of SAH.

The initial cerebral angiogram failed to depict the cause of SAH in seven (35%) of 20 patients. Six cDAVFs arose from a radicular branch of the right vertebral artery (86%), and one arose from the left. Twelve initial angiograms were reported as being positive. The arterial supply was from the right in five cases.

Discussion

Kendall and Logue (27) pioneered the classification of spinal vascular malformations, dividing them into dural and intradural types. Spinal DAVFs are acquired abnormal communications between the arterial and venous system within the dura of the prox-

imal dorsal nerve root and adjacent spinal dura. The arterial supply is usually from a radiculomeningeal branch of the segmental artery. Usually, a single fistula drains via a radicular vein into the coronal venous plexus and, to a lesser extent, into the epidural and paravertebral veins.

Tadie et al (28) performed microangiography on normal radicular veins and demonstrated a constriction at the point at which they cross the dura. The constriction acts as a valve that prevents the transmission of pressure into the valveless coronal venous plexus. Therefore, for shunt surgery to occur, the fistula must be located at this point or an intradural point. Benhaiem et al (29) confirmed these findings histologically. Once a fistula has formed, the increased medullary venous pressure is transmitted through the coronal venous plexus into the cord via the radial veins. The arteriovenous pressure gradient and spinal blood flow are reduced because of increased intramedullary pressure. Venous hypertension results in myelopathy (30). Merland et al (3) and Hassler et al (31) expanded on this theory and described additional, insufficient venous drainage in symptomatic spinal DAVFs due to a lack of normal draining radicular veins. The myelopathy in DAVFs is pathologically referred to as subacute necrotizing myelopathy. This is thought to be the same as the condition first described by Foix and Alajouanine (32) and subsequently pathologically confirmed by Hurst et al (33).

DAVFs in the cervical spine and at the craniocervical junction are rare. Although the age distribution of patients with these DAVFs is similar to those with lesions in the more common thoracolumbar region (1, 2), the heavily weighted male predominance is less obvious, with a ratio of 2:1 to 3:1 versus 7.5:1 (2). cDAVFs have a wide variation in presentation that includes myelopathy, radiculopathy, cranial nerve palsies, and SAH. Two case reports not included in this review describe presentations with occipitalgia (34) and transient ischemic attack (35).

For the purpose of statistical comparison, upper cDAVFs were considered as those arising between the foramen magnum and the C2 inclusive. The cervical level was not statistically correlated with a presentation of hemorrhage. The presence of a venous pouch and/or varix is not described in the thoracolumbar region (1), and its presence is associated with SAH at presentation.

cDAVFs appear more often to have alternative routes of drainage, including intracranial ones. Drainage can also be purely epidural, as in both our cases. This finding may be explained by the DAVF lying proximal to that portion of the medullary vein spanning the dura. Some neurointerventionalists question the ability of DAVFs to drain epidurally; however, in one of our cases, a DAVF with epidural drainage was surgically confirmed. Seven other cases from six series are described as having epidural drainage with or without intradural drainage (5, 9, 12, 14, 15, 18). Three of the seven patients were surgically treated, and DAVFs were confirmed intraoperatively. Two of

these patients presented with SAH. One patient receiving endovascular treatment (5) had epidural venous drainage and presented with SAH. Myelopathy is described in three cases with pure epidural drainage (5, 14, 15), which is presumed to be based on mass effect. This discussion illustrates the fact that a DAVF may have epidural drainage. The presence of superior drainage in the cervical region is indicative of abnormal venous drainage, which is a common finding in DAVF. This is thought to result from thrombosis of the normal pathways of venous return, which is usually caudal, above the level of the heart. Superior and/or intracranial drainage of a cDAVF is significantly associated with the presentation with SAH.

There is no significant difference in the level of origin of the supplying radicular artery or in the side of supply in the SAH group compared with the non-SAH group. Approximately 63% of cDAVFs arose from the right vertebral artery. Of seven patients with SAH for whom initial angiograms were negative, six had cDAVFs supplied from the right vertebral artery. This observation has implications for angiographically negative SAH, as the left vertebral artery is most frequently dominant and the single vertebral artery selectively injected if sufficient reflux down the right vertebral artery is achieved to opacify the right posterior inferior cerebellar artery. It is noteworthy that all cDAVFs discovered at initial angiography (12 of 20) occurred at the craniocervical level, which is usually included in the field for cerebral angiography of the posterior circulation. However, 40% were missed, and these were most often supplied from the right vertebral artery. We advise opacification of both vertebral arteries, with views of the neck in cases of intracranial SAH with negative cerebral angiograms. Failure to identify a cDAVF after injection of both vertebral arteries warrants a thorough evaluation of the thyrocervical, costocervical, and ascending pharyngeal arteries to look for another segmental cervical supply.

Our sample size was not large enough to detect moderate or small differences between the SAH and non-SAH groups. Therefore, any lack of significant difference should not be seen as evidence of similarity. However, in light of the rarity of DAVFs, our review is important because it identified two features—intracranial drainage and venous varix—that are present in cDAVFs in patients presenting with subarachnoid bleed. Whether these factors predict the risk of hemorrhage cannot be proved unless a cohort of incidentally discovered, asymptomatic cDAVFs are followed with angiography until they manifest clinically. Both of these features are rare in thoracic DAVFs, and this might possibly explain the increased risk of bleeding in cDAVFs.

We have highlighted the importance of selectively injecting both vertebral arteries, as twice as many cDAVFs are supplied from the right than from the left. When an angiogram is negative for SAH, a thorough evaluation of the cervical segmental arterial supply should be performed to exclude a cDAVF.

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