Spinal dural arteriovenous fistula: correlation between radiological and clinical findings

Clinical article

PEGGY P. W. YEN, M.D.,1 KRISTA C. RITCHIE, M.A., PH.D.,2,3 AND JAI JAI SHIVA SHANKAR, M.D., D.M., M.SC.4

1Department of Diagnostic Radiology, Queen Elizabeth II Health Sciences Centre, Dalhousie University; 2Interdisciplinary Research, Dr. Richard B. Goldbloom Research and Clinical Care Pavilion, IWK Health Centre; 3Department of Community Health and Epidemiology, Faculty of Medicine, Dalhousie University; and 4Department of Diagnostic Radiology, Division of Neuroradiology, Queen Elizabeth II Health Sciences Centre, Dalhousie University, Halifax, Nova Scotia, Canada

Object. The pathophysiology of spinal dural arteriovenous fistulas (SDAVFs) results in perimedullary venous congestion and in turn central cord congestion. Clinically, this presents with progressive neurological dysfunctions that, if diagnosed in a timely fashion, can be at least halted and in part reversed.

In SDAVFs, imaging features on MRI and digital subtraction angiography (DSA) have not been studied in conjunction with clinical findings. The primary purpose of the present study was to test if severity of clinical presentation varies in relation to imaging.

Methods. This retrospective cohort study identified 12 patients treated for SDAVF at the authors’ institution. The extent of venous congestion and cord edema was quantified by the number of vertebral levels shown to be affected on DSA and MRI. A modified Aminoff-Logue Scale (ALS) score was assigned at the time of diagnosis and again after definitive therapy. The patients were divided into one of two groups: those with venous congestion < 7 and ≥ 7 vertebral levels seen on DSA and MRI and with central cord edema < 6 and ≥ 6 levels. A t-test was used to assess for a difference in the presenting ALS score between the groups.

Results. Patients with ≥ 7 levels of venous congestion reported greater functional disability (DSA: p ≤ 0.001, Cohen’s d = 0.509; and MRI: p ≤ 0.001, d = 0.632). Patients with a greater extent of cord edema also reported worse functional disability (p ≤ 0.001, d = 2.31). There was a strong linear correlation between the post- and pretreatment ALS scores (R² = 0.86) for those with successful interventions (n = 9).

Conclusions. In patients with an SDAVF, the severity of the neurological dysfunction may be predicted by the extent of DSA- and MRI-documented venous congestion and cord edema. There was a strong positive relationship between initial and posttreatment neurological dysfunction.

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Key Words • dural arteriovenous fistula • magnetic resonance imaging • digital subtraction angiography • embolization • spinal cord disease • vascular disorders

Spinal dural arteriovenous fistula (SDAVF) is a rare, yet the most frequently acquired, spinal vascular malformation that can devastatingly impair spinal cord function.9,12 The presenting clinical symptoms are nonspecific, and thus it is often initially misdiagnosed, with a reported diagnostic delay of 15–23 months, at which time 78% of patients have been reported to have severe lower-limb weakness and 19% to be wheelchair bound.17 Early diagnosis is very important since the symptoms can be reversed if treated prior to irreversible cord ischemia or infarction.

Serial imaging provides the definitive diagnosis of SDAVF.11 The typical order of imaging modalities starts with whole-spine MRI to establish the diagnosis of an SDAVF and to rule out other causes of neuropathy. Characteristic features on spinal MRI include multilevel centromedullary cord T2 hyperintensity signifying spinal cord edema, and this is sometimes accompanied by a hypointense rim, which is postulated to represent deoxygenated blood within the dilated capillary vessels.3 In later stages of the disease, the cord can become atrophic due to chronic hypoxia. The abnormal perimedullary vessels
are congested and will appear dilated and tortuous. On T2-weighted MRI sequences, this can be visualized as continuous prominent flow voids in the extramedullary and intradural space (Figs. 1 and 2). None of these features actually localizes the site of fistula. Spinal digital subtraction angiography (DSA) helps to define the fistula site, the feeding artery, and drainage pattern and can concurrently assist in the therapeutic embolization of the fistula.

Early in the course of the disease, patients often present with nonspecific symptoms including low-back pain, unilateral or bilateral lower-limb paresthesias and weakness, or gait disturbances. With a decreased arteriovenous gradient, there is decreased neural tissue perfusion, resulting in neuronal depopulation, gliosis, and lastly necrosis of white or gray matter. Due to the caudal distribution of the venous congestion, the conus medullaris is often affected and progresses in a caudocranial direction. Hence, regardless of the fistula level, patients often will present with common complaints of sensory loss, neurogenic bladder, and lower-extremity myelopathy.

Due to the final common pathogenetic pathway of venous congestion, the imaging features have not been thought to be indicative of clinical severity. The primary purpose of the present study was to analyze whether the severity of functional disability was greater for patients with extensive imaging findings. This correlation is important because early reversal of the venous congestion can arrest symptom progression, and if no definitive histological lesions are already present, early intervention has been shown to reverse paraplegia within 6 months postintervention. A secondary goal of the study was to examine the relationship between functional status before and after treatment and to test whether functional status improved after treatment.

Methods

We performed a retrospective review of all patients with the diagnosis of SDAVF in the past 10 years (2003–2013) in our interventional neuroradiology database. The study was reviewed and approved by our institutional ethics committee. The diagnosis of SDAVF was confirmed by serial imaging, including MRI evaluation of the entire spine and spinal DSA. The clinical information including age, sex, symptoms, SDAVF location, treatment, and symptoms at approximately 6 months posttherapy were obtained through a retrospective chart review for each patient.

Twelve patients were identified—8 men and 4 women whose mean age (± SD) was 53 ± 15 years (range 22–71 years). The fistulas were located at the cervical (n = 3), thoracic (n = 5), lumbar (n = 2), and sacral (n = 2) levels. Eight patients underwent definitive glue embolization, 2 underwent surgical therapy, 1 required a repeat glue emb-
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bolization, and 1 underwent surgical intervention after a failed embolization. In 2 patients intraoperative complication occurred, resulting in cord infarct in one and non-target glue embolization of the left vertebral artery and subsequent brainstem and cerebellar infarction in the other.

The clinical severity at presentation and at 6 months posttreatment was documented and quantified in accordance with the modified Aminoff-Logue Scale (ALS) based on the gait and micturition abnormalities.2 The ALS assigns a score to gait and micturition functions, with 0 representing normal status. Greater weighting is given to mobility, and the higher the score, the more severe the functional disability. For gait a maximal gait score of 5 is assigned when the patient is wheelchair bound, and for micturition a maximal score of 3 is given to patients experiencing total incontinence or persistent urinary retention.

Image Analysis

Complete review of the MRI and DSA exams was performed by the investigators (J.S. and P.Y.), who were blinded to the clinical information of the patient. The T2-weighted images were analyzed for the presence of central cord hyperintensity and perimedullary flow voids independent of the presence of the other. The severity of these imaging findings was quantified by counting the vertebral levels at which the abnormalities were present. Image analysis was performed for only 11 patients since the preoperative MRI of one patient was conducted in an outside institution, and the studies could not be obtained for our review.

The DSA images were assessed for the presence of tortuous and dilated perimedullary veins. Where present, the extent was again quantified by the number of vertebral bodies the abnormality traversed in both the cranial and caudal directions. This is relevant in cases in which the enlarged draining vein traveled caudally before ascending cranially. Because of the retrospective nature of the review, the vertebral-level image coverage was limited, depending on the operator’s preference. The cervical region was usually not imaged, thereby underestimating the affected levels. Only 3 patients had complete evaluation of the spinal cord. For this reason, the maximum number of levels visible were recorded and used in the statistical analysis.

Statistical Analysis

The data were arranged and analyzed within Excel (Microsoft Office 2007). On post hoc analysis, the patients were divided into 2 groups, those with ≥ 7 and < 7 vertebral levels of dilated intrathecal veins on both MRI and DSA. Patients were also divided into those with ≥ 6 and < 6 vertebral levels of cord edema. The classifications into the 2 numerical groups were determined by the overall data distribution (See Table 1 for full data set). An attempt was made to maintain a similar cutoff level for all 3 variables. The difference in the means of ALS scores between the 2 groups was calculated using 2-tailed Student t-tests. Correlation analysis between the functional disability at presentation and 6 months after the final therapy was performed by fitting a linear regression and the coefficient of determination (R²).

Due to the small sample size, a standardized effect size is determined by calculating the Cohen’s d value for the t-test results. The Cohen’s d value gives the magnitude of difference between the 2 groups for a given variable and indicates the distance between the 2 means, taking into account the variance in standard deviation units. This is considered complementary to the reporting of statistical significance and allows for comparison between different studies. For Cohen’s d, by convention, a value of less than 0.2 equates to a small effect, 0.5 to a moderate effect, 0.8 to a large effect, and greater than 1 to an extremely large effect size.5

Results

The mean number of vertebral levels of venous congestion on DSA and MRI was 8.0 ± 3.4 (median 7) and 10.2 ± 8.1 (median = 9), respectively. The mean number of vertebral levels affected by central cord edema was 5.8 ± 3.5 (median 6). Patients with a greater number of levels affected by venous congestion and cord edema had higher total ALS scores. The presenting functional disability ranged from 1 to 8. Three patients had no micturition symptoms, but all had at least unilateral or bilateral lower-extremity paresthesia. Three patients were wheelchair dependent and 2 required a walker for ambulation. Two of the most severely ambulatory-restricted patients also had complete urinary incontinence requiring regular catheterization. The ALS scores for patients with ≥ 7 vertebral levels of venous congestion were similar whether diagnosed by DSA or MRI (scores of 4.7 ± 2.4 and 4.3 ± 2.7, respectively). These scores were significantly higher than those with venous congestion affecting < 7 vertebral levels (3.4 ± 2.7 [p ≤ 0.001] and 2.7 ± 2.9 [p ≤ 0.001], respectively). Patients with cord edema affecting ≥ 6 vertebral levels had a mean ALS score of 5.3 ± 1.9 (Fig. 1) whereas those with cord edema affecting < 6 vertebral levels had a mean score of 1.8 ± 1.0 (p ≤ 0.001) (Table 2; Fig. 2). After accounting for sample size, there remained a moderate difference between the DSA (d = 0.509) or MRI (d = 0.632) groups and the ALS at presentation. There was an extremely large effect between the cord edema groups (d = 2.31) and the ALS at presentation.

One patient had operative complications resulting in death due to brainstem and cerebellar infarction, and another patient had severe morbidity due to an intraoperative cord infarct. The remaining 10 patients had a relatively uneventful postoperative recovery and were followed up closely for functional status. There was a strong linear correlation (R² = 0.86) between the severity of functional disability at presentation and that at approximately 6 months after treatment. The posttherapy ALS score could be predicted using the linear predictor function y = 1.21x − 3.0, where x equals the ALS score at presentation (Fig. 3). Intuitively, this conveys a better prognosis for those with less neurological dysfunction at presentation, underscoring the importance of an early diagnosis and treatment.
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex, Location</th>
<th>No. of Vertebral Levels</th>
<th>Venous Congestion</th>
<th>Cord Edema</th>
<th>Pretherapy ALS Score</th>
<th>Symptoms</th>
<th>Therapy</th>
<th>Posttherapy ALS Score</th>
</tr>
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<tr>
<td></td>
<td></td>
<td></td>
<td>DSA</td>
<td>T2WI</td>
<td>T2WI</td>
<td>Mict</td>
<td>Amb</td>
<td>Total</td>
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<td>1</td>
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<td>&gt;10</td>
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<td>NP</td>
<td>NP</td>
<td>3</td>
<td>5</td>
<td>8</td>
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<tr>
<td>2</td>
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<td>&gt;6</td>
<td>18</td>
<td>0</td>
<td>1</td>
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<td>NP</td>
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<td>0</td>
<td>1</td>
<td>rt weakness</td>
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<td>7</td>
<td>9</td>
<td>2</td>
<td>5</td>
<td>7</td>
<td>requires wheelchair; frequent incontinence</td>
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<tr>
<td>6</td>
<td>31, M, rt T-11</td>
<td>&gt;5</td>
<td>9</td>
<td>4</td>
<td>2</td>
<td>1</td>
<td>3</td>
<td>bilat weakness &amp; total incontinence</td>
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<tr>
<td>7</td>
<td>71, F, rt L3–4</td>
<td>&gt;13</td>
<td>17</td>
<td>6</td>
<td>3</td>
<td>4</td>
<td>7</td>
<td>requires walker; total incontinence</td>
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<tr>
<td>8</td>
<td>57, F, lt S-1</td>
<td>&gt;10</td>
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<td>10</td>
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<td>3</td>
<td>3</td>
<td>uses walker</td>
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<tr>
<td>9</td>
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<td>&gt;10</td>
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<td>5</td>
<td>6</td>
<td>wheelchair dependent; total incontinence</td>
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<td>68, M, lt L-2</td>
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<td>0</td>
<td>6</td>
<td>2</td>
<td>4</td>
<td>6</td>
<td>bilat weakness; total incontinence</td>
</tr>
</tbody>
</table>

* Amb = ambulation; embo = glue embolization; lam = laminectomy; Mict = micturition; NP = MRI not performed; surg = surgery; T2WI = T2-weighted image.
† Intraoperative cord infarction.
‡ Glue into left vertebral artery resulted in cerebellar and brainstem infarct.
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TABLE 2: Degree of functional disability at the time of diagnosis based on the extent of venous congestion or cord edema

<table>
<thead>
<tr>
<th>Modality</th>
<th>No. of Cases</th>
<th>ALS Score (mean ± SD)</th>
<th>p Value</th>
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</thead>
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<tr>
<td>venous congestion</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>DSA</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>≥7 levels</td>
<td>7</td>
<td>4.7 ± 2.4</td>
<td>0.004</td>
</tr>
<tr>
<td>&lt;7 levels</td>
<td>5</td>
<td>3.4 ± 2.7</td>
<td></td>
</tr>
<tr>
<td>MRI</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>≥7 levels</td>
<td>8</td>
<td>4.3 ± 2.1</td>
<td>0.004</td>
</tr>
<tr>
<td>&lt;7 levels</td>
<td>3</td>
<td>2.7 ± 2.9</td>
<td></td>
</tr>
<tr>
<td>cord edema</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>≥6</td>
<td>6</td>
<td>5.3 ± 1.9</td>
<td>0.003</td>
</tr>
<tr>
<td>&lt;6</td>
<td>4</td>
<td>1.8 ± 1.0</td>
<td></td>
</tr>
</tbody>
</table>

Discussion

Spinal dural arteriovenous fistulas are rare lesions, and early diagnosis is crucial because progressive myelopathy is potentially reversible. From an imaging standpoint, it is well established that the complete spinal MRI examination is diagnostic for the presence of an SDAVF, but now we have shown that the extent of the imaging abnormalities may have prognostic value as well. Our small series demonstrated that the craniocaudal extent of perimedullary venous congestion may be indicative of the severity of the clinical symptoms. Intramedullary edema develops because the intramedullary veins and the radicular veins share a common venous outflow. Chronic intramedullary congestion leads to hypoxia and ensuing progressive myelopathy.

When the 2 cases of intraoperative complication are excluded from our series, the progression of disease was at least halted if not partially reversed in 9 of 9 patients when prompt occlusion of the fistula was achieved. The degree of symptom regression was dependent on the pretreatment clinical status. A linear relationship exists in which the more severe the presenting symptoms, the greater is the residual functional disability following treatment (Fig. 3). This is likely related to the permanent spinal myelomalacia due to the chronic hypoxia.

Hetts et al.² have shown a similar relationship in which there was a correlation between worsening spinal cord dysfunction and greater extent of intrathecal venous drainage. In the present report, we have also shown that this relationship can be demonstrated using either MRI or DSA and furthermore that central cord edema is predictive of functional disability. The correlation between severity of disease and imaging characteristics is greater when the cutoff for the levels of venous congestion is set at ≥7 vertebral levels rather than the ≥10 set by Hetts et al. This may reflect a greater confidence in correlating imaging features with the clinical presentation and ultimately in predicting reversibility of the symptoms when a less stringent cutoff level (in our case a cutoff of 7 vertebral levels) is used as the threshold.

Due to the rarity of this disease process, the sample size was small and further reduction in number was necessary because of patients' intraoperative complications. Despite the small sample size, there is at least a moderate to a large effect size when the sample size is accounted for in the Cohen's d calculation. The findings of our study, in conjunction with those of Hetts et al.,² suggest the potential for imaging features indicating the clinical severity of this rare disease.

Our results have to be further confirmed in a larger series in a prospective fashion. In addition, because of the retrospective nature of the study design, complete assessment of the perimedullary venous congestion was not performed consistently, and the exact extent of involvement could not be determined. However, by dividing the number of affected levels into the 2 categories, some of the uncertainty could be overcome. In light of the likely correlation between venous congestion and functional status, it may be worthwhile in the future to document the extent of venous congestion on spinal DSA.

Conclusions

The severity of neurological dysfunction in SDAVF patients may be correlated with the extent of imaging abnormalities found on DSA and MRI. The patients with a greater craniocaudal extent of venous congestion or central cord edema exhibited severe neurological dysfunction. The strong positive relationship between initial and posttreatment neurological dysfunction emphasizes the need for early treatment in patients with SDAVF.

Disclosure

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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Author contributions to the study and manuscript preparation include the following. Conception and design: Shankar, Yen. Acquisition of data: Yen. Analysis and interpretation of data: Shankar, Yen. Drafting the article: Yen. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Shankar. Statistical analysis: Yen, Ritchie. Administrative/technical/material support: Shankar, Yen.

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P. P. W. Yen, K. C. Ritchie, and J. J. S. Shankar