Dural arteriovenous fistulas presenting with brainstem dysfunction: diagnosis and surgical treatment

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A cerebral dural arteriovenous fistula (DAVF) is an acquired abnormal arterial-to-venous connection within the leaves of the intracranial dura with a wide range of clinical presentations and natural history. The Cognard classification correlates venous drainage patterns with neurological course, identifying 5 DAVF types with increasing rates of symptomatic presentation. A spinal DAVF occurs when a radicular artery makes a direct anomalous shunt with a radicular vein within the dural leaflets of the nerve root sleeve. A cervical DAVF is a rare entity, as most spinal DAVFs present as thoracolumbar lesions with myelopathy. In this paper the authors present 2 patients presenting initially with brainstem dysfunction rather than myelopathy secondary to craniocervical DAVF. The literature is then reviewed for similar rare aggressive DAVFs at the craniocervical junction presenting with brainstem symptomatology.

Case Reports

Case 1

This 44-year-old woman presented with acute onset of altered mental status, right hemiparesis, and limb dyscoordination. Subsequently, she became hypopneic when asleep and was transferred to a nursing home after undergoing a tracheostomy for central hypoventilation; the final diagnosis of her condition was “brainstem stroke” (Fig. 1). She subsequently developed episodes of abnormal limb movement, believed to be exacerbation of her brainstem ischemia based on neurological assessment.
Retrospective evaluation of her initial workup, including CT angiography, revealed prominent skull-base/high-cervical vasculature (Fig. 2). Formal angiography revealed a DAVF. The major arterial supply was the left MMA, with a minor arterial supply from a dural branch of the left vertebral artery (Fig. 3). The fistula connected to the left superior petrosal sinus with drainage via a perimesencephalic vein through the prepontine venous system and into the perimedullary venous plexus.

Due to the persistent and progressive symptoms of her brainstem dysfunction, intervention was considered necessary. Open surgical disconnection of the fistula through a retromastoid craniotomy was performed. Two arterialized veins were found originating from the dura along the tentorial-petrous junction and were clip ligated; 1 traveled along the anterior brainstem and was ligated using a clip just anterior to the trigeminal nerve (Fig. 4.). She recovered well from surgery. Postoperatively, she progressively demonstrated improved right-sided motor function, albeit with residual paresis. Her chronic central hypoventilation was subsequently managed with phrenic nerve stimulation. Due to her stimulator, she could not undergo postoperative MRI evaluation.
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Case 2

This 54-year-old woman presented with acute onset of left hemiparesis and diplopia. Intravenous thrombolytics led to minimal neurological improvement. Magnetic resonance imaging suggested ischemic infarction of the right inferior pons (Fig. 5). This infarction was presumed to be due to small vessel disease as no vertebrobasilar or cardiac source was identified. After discharge to rehabilitation, she suffered from another neurological decline with worsening of her left-side weakness and development of a new right-side weakness. New imaging studies revealed enlargement of her right pontine lesion and a new left pontine ischemic abnormality (Fig. 6). Cervical and cerebral angiography demonstrated a DAVF arising from the right vertebral artery (Fig. 7) originating from the right C-3 segmental branch of the artery, with medullary venous outflow draining in a cephalad direction along the cervical spinal cord and brainstem.

A second stroke workup found no underlying cause for her progressive brainstem ischemia except for her cervical DAVF. The patient underwent a cervical laminectomy and DAVF disconnection. Intraoperative findings noted 3 fistulous vessels arising anteriorly adjacent to the C-2 nerve root, all of which were coagulated and cut (Fig. 8). She experienced progressive right-sided improvement in her strength after surgery. Follow-up 6-month MRI after surgery demonstrated no further areas of ischemia, decreased T2-signal change (indicating resolution of venous congestion), and small areas of pontine encephalomalacia (Fig. 9).

Discussion

Case reports of DAVFs presenting with primarily brainstem ischemia are very rare. Our literature search followed by a secondary review of the associated references in the primary sources revealed only 4 reported Cognard Type V DAVFs presenting as brainstem ischemia in the English literature, with a fifth case in the Japanese literature. Only 1 other cervical DAVF presenting with brainstem ischemia was found. These reported cases are summarized in Table 1.

Dural arteriovenous fistulas adjacent to the cranio-cervical region most often present with progressive myelopathy related to spinal cord ischemia/venous hypertension. In this paper we report 2 such cases that atypically presented with brainstem ischemia. Although rare, the progressive natural history of these lesions and their resolution with intervention demonstrates the importance of detection in a timely fashion. Our first patient was initially diagnosed with a brainstem stroke, and no further workup was considered necessary. Our report emphasizes the need to further evaluate brainstem stroke of unknown origin through more comprehensive vascular studies, including cerebral angiograms.

The primary mechanism responsible for brainstem dysfunction among the patients noted above is believed to be venous hypertension secondary to arterial pressure via the fistula. This mechanism has previously been suggested to cause potentially reversible venous congestion. In the first case, increased T2 signal changes within the brainstem without analogous diffusion restriction on
diffusion-weighted imaging/ADC mapping was consistent with previously described venous congestion that has been documented to resolve after fistulous disconnection.\(^6\) The typical ascending course is related to venous congestion. This congestion is observed initially in the anterior spinal vein, ascending via the anterior medullary vein to the anterior pontomesencephalic vein, worsened by vascular “steal” from normal spinal blood supply.\(^8\) However, due to the primarily external carotid arterial source, a steal phenomenon involving the spinal cord vessels was unlikely. This possibly explains the relative tolerance of the spinal cord to venous hypertension and resultant lack of myelopathy that usually dominates the initial presentation of Cognard Type V fistulas.

In the second case, however, initial MRI was more suggestive of ischemic disease, with increased diffusion-weighted signal and corresponding low ADC signal. In spite of this fact, a thorough embolic workup and empirical treatment with antiplatelet agents were unsuccessful. The patient’s ischemic lesions and symptoms subsequently progressed. The presence of restricted diffusion combined with rostral vertebral blood supply suggests the possibility of a steal phenomenon, which could result in ischemia to tissue already at risk due to concomitant venous hypertension. The only prior case report of a cervical DAVF presenting with brainstem dysfunction did
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As shown by the 2 cases in this report, cranio cervical DAVF should be considered in the differential diagnosis of ischemic brainstem dysfunction of unknown origin. Prompt diagnosis and treatment are crucial to provide the patient with the best chance at improvement.

Conclusions

Our 2 cases demonstrate that although previous reports largely relied on endovascular embolization occlusion techniques, open surgical disconnection is feasible, safe, and efficacious for both cerebral and cervical lesions. Interestingly, in all cases with reported outcomes, the progressive neurological decline ceased, and prompt neurological improvement occurred (albeit mild in some instances) after fistulous interruption. Prior cases have demonstrated reversibility of the venous congestion-related dysfunction, emphasizing the importance of timely recognition and treatment, which mandates inclusion of a cranio cervical DAVF in the differential diagnosis of new brainstem dysfunction. Our 2 cases demonstrate that although previous reports largely relied on endovascular embolization occlusion techniques, open surgical disconnection is feasible, safe, and efficacious for both cerebral and cervical lesions. Interestingly, in all cases with reported outcomes, the progressive neurological decline ceased, and prompt neurological improvement occurred (albeit mild in some instances) after fistulous interruption. Prior cases have demonstrated reversibility of the venous congestion-related dysfunction, emphasizing the importance of timely recognition and treatment, which mandates inclusion of a cranio cervical DAVF in the differential diagnosis of new brainstem dysfunction.

T2 hyperintensity without associated diffusion restriction may be more predictive of prompt recovery than findings of diffusion restriction, possibly due to an alternative mechanism of more reversible injury.

TABLE 1: Reported cases of cranio cervical DAVF presenting with brainstem symptomatology

<table>
<thead>
<tr>
<th>Type of DAVF</th>
<th>Authors &amp; Year</th>
<th>Presenting Symptoms</th>
<th>Location of Dysfunction</th>
<th>Arterial Source</th>
<th>Venous Sinus Drainage</th>
<th>Treatment</th>
<th>Neurological Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>cerebral</td>
<td>Li et al., 2004</td>
<td>vertigo, nausea &amp; vomiting, paraparesis</td>
<td>pontomedullary</td>
<td>MMA, OA</td>
<td>transverse</td>
<td>coil embolization</td>
<td>improved</td>
</tr>
<tr>
<td></td>
<td>Oishi et al., 2005†</td>
<td>disturbance of brainstem function</td>
<td>medulla</td>
<td>unknown</td>
<td>superior petrosal</td>
<td>coil embolization</td>
<td>unknown</td>
</tr>
<tr>
<td></td>
<td>Satoh et al., 2005</td>
<td>vertigo, nausea &amp; vomiting, dysphagia, rt motor &amp; sensory changes</td>
<td>medulla</td>
<td>MMA, OA, APA, MHT, PMA</td>
<td>transverse-sigmoid</td>
<td>coil embolization</td>
<td>improved</td>
</tr>
<tr>
<td></td>
<td>Lagares et al., 2007</td>
<td>vertigo, quadriaparesis, dysphagia, respiratory insufficiency</td>
<td>pontomedullary</td>
<td>OA, PMA</td>
<td>transverse</td>
<td>surgical disconnection</td>
<td>improved</td>
</tr>
<tr>
<td></td>
<td>Sugiura et al., 2009</td>
<td>nausea &amp; vomiting, ataxia, nystagmus</td>
<td>pontomedullary</td>
<td>OA</td>
<td>sigmoid</td>
<td>coil embolization</td>
<td>improved</td>
</tr>
<tr>
<td></td>
<td>present study, Case 1</td>
<td>confusion, hemiparesis, ataxia</td>
<td>pontomedullary</td>
<td>MMA, PMA</td>
<td>superior petrosal</td>
<td>surgical disconnection</td>
<td>improved</td>
</tr>
<tr>
<td>cervical</td>
<td>Terao et al., 2006</td>
<td>vertigo, orthostasis, urinary retention, paraparesis</td>
<td>medulla</td>
<td>C-6 segmental artery off VA</td>
<td>spinal perimedullary vein</td>
<td>coil embolization, surgical disconnection</td>
<td>improved</td>
</tr>
<tr>
<td></td>
<td>present study, Case 2</td>
<td>hemiparesis, diplopia</td>
<td>pons</td>
<td>C-3 segmental artery off VA</td>
<td>spinal perimedullary vein</td>
<td>surgical disconnection</td>
<td>improved</td>
</tr>
</tbody>
</table>

* APA = ascending pharyngeal artery; MHT = meningohypophyseal trunk; OA = occipital artery; PMA = posterior meningeal artery; VA = vertebral artery.
† Only available in Japanese literature.
Disclosure

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

Author contributions to the study and manuscript preparation include the following. Conception and design: Cohen-Gadol, Kulwin. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors.

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