Cervical spine dural arteriovenous fistula presenting with congestive myelopathy of the conus

Case report

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Spinal dural arteriovenous fistulas (DAVFs), which are also classified as Type I spinal AVMs or are called the acquired type, are the most common spinal vascular malformations, accounting for nearly 70% of such lesions.10 They are typically located at the thoracolumbar region and are found predominantly in middle-aged to elderly men. Cervical locations are rare and can be further subdivided into upper and lower levels; the former being at the level of the foramen magnum and the C1–2 level, with the latter in the levels below C-2. The upper or cranio cervical locations may present either with venous congestion of the cervical spine or SAH, depending on the venous drainage pattern.1,25 Lower cervical spine DAVFs are extremely rare, with only 8 cases previously reported in the literature, and with varying clinical presentations.2,4,8,15,25,26

We present a case of a cervical DAVF located at the C-5 level that presented with congestive venous edema at the lower thoracic cord and conus medullaris.

Case Report

History and Examination. This 51-year-old man presented with progressive weakness of both legs that had lasted for 4 months. He had urinary incontinence and constipation, with numbness of his buttocks. On admission, physical examination revealed a spastic gait with Grade III paraparesis (which was pronounced on the left side) according to the modified McCormick scale.14 Decreased pinprick sensation from the L-1 level was noted, with a loose sphincter tone.

Neuroimaging. Admission T2-weighted MR imaging revealed hypersignal of the spinal cord from T-7 down to the conus at the T-12 level, with abnormal dilated perimedullary vessels at the dorsal aspect of the spine, which were present from the conus to the lower cervical spine. There was mild degenerative disc dis-
ease. Both clinical and imaging findings were suggestive of a spinal vascular malformation, and complete spinal digital subtraction angiography was performed, starting from the lower thoracic level. The ASA was constituted by radiculomedullary vessels that arose from the left T-7 intercostal artery and the left L-2 lumbar artery. There was a markedly delayed venous phase at the lower thoracic level (>20 seconds). After the thoracic and lumbar segment arteries were selectively injected without localizing the shunt, the cervical arteries were checked. The left VA angiogram revealed a slow-flow AVS, supplied by a radicular artery arising from the left C-5 level and draining into descending PMVs, that could be opacified in the late phases to the level of the conus (Fig. 1). The right VA and the remaining cervical arteries were normal. Radiculomedullary arteries supplying the ASA were identified at bilateral C-6 levels from both VAs. There was no contribution to the shunt from these arteries. The diagnosis of a cervical DAVF arising from the C-5 level was made.

Operation and Postoperative Course. Subsequently, transarterial N-butyl-cyanoacrylate glue embolization through the left C-5 radicular artery via a 1.2 Fr Magic microcatheter was performed successfully; the glue cast closed the shunt and the proximal part of the primary venous drainage, resulting in complete obliteration of the DAVF. Clinical follow-up performed at 2 months showed nearly complete resolution of the paraparesis, with only a mild left foot drop remaining. Bowel and bladder symptoms were also improved. Follow-up MR imaging confirmed the complete obliteration of the shunt. No abnormal vessels were seen in the perimedullary area, and the congestive edema of the spinal cord was markedly decreased (Fig. 2).

Discussion

Spinal DAVFs are supplied by radiculomeningeal arteries and drain into radicular veins, which connect centripetally with either ascending or descending PMVs. The AVS is located inside the dura mater close to the spinal nerve root, where the arterial blood enters the radicular vein (where the latter passes the dura). The increase in spinal venous pressure diminishes the arteriovenous pressure gradient and leads to a decreased drainage of normal spinal veins and a venous congestion, with intramedullary congestive edema. This in turn leads to chronic hypoxia and progressive myelopathy. With the exception of foramen magnum spinal DAVF, hemorrhagic presentation is never encountered.

Occasionally, the angiographic characteristics described above may be difficult to differentiate from those of other vascular lesions: that is, radicular AVMs, epidural AVSs, or perimedullary AVFs. Radicular AVMs or AVMs of the nerves usually have a conglomerate of abnormal vessels that form a nidus surrounding the nerve root, whereas spinal DAVFs will have an apparent shunt zone, with radicular feeding vessels converging into the same draining vein. Clinically, patients suffering from a radicular AVM often present with radicular pain and only rarely show signs of congestive venous myelopathy. Epidural AVSs are located in the epidural space and recruit the arterial supply of the vertebral body and its surrounding structures, with drainage directed into the epidural plexus and not into PMVs. Symptoms are
related to compression of the adjacent nerve root or spinal cord rather than venous congestion, unless exceedingly rarely, when perimedullary reflux occurs due to a failure of the antireflux mechanism from the epidural venous space into the PMVs.\(^9,17,19\) Perimedullary AVFs are usually high-flow shunts that are located at the surface of the spinal cord and that are necessarily supplied by arteries that also supply the cord; that is, either through radiculomedullary arteries forming the ASA or radiculopial arteries via the posterolateral spinal arteries.\(^10\) In high-flow fistulas, venous pouches may be encountered, especially in young patients with hereditary hemorrhagic telangiectasia.\(^11\) Symptoms may be similar to those of spinal DAVFs; however, there is no gender predominance in perimedullary fistulas, and they tend to occur in younger patients compared with spinal DAVF.

In the literature, which is summarized in Table 1, there are 8 patients reported in whom a cervical DA VF was considered as the underlying disease. Due to the above-mentioned considerations concerning potential differential diagnoses, some of these cases may in fact be diagnosed as other types of shunts. The cases reported by Cahan et al., Willinsky et al., Yamada et al., and Kohno et al.\(^8\) demonstrate drainage of the AVSs directly into large epidural venous pouches. There is no shunt into radicular veins or even PMVs, and the clinical presenta-

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Level</th>
<th>Age (yrs), Sex</th>
<th>Presentation</th>
<th>Venous Drainage</th>
<th>Possible Dx</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cahan et al., 1987</td>
<td>rt C-5</td>
<td>19, M</td>
<td>SAH</td>
<td>CVP, epidural veins</td>
<td>epidural AVSs</td>
</tr>
<tr>
<td></td>
<td>C4–5</td>
<td>51, F</td>
<td>radiculopathy, bruit</td>
<td>epidural veins</td>
<td>epidural AVSs</td>
</tr>
<tr>
<td>Willinsky et al., 1990</td>
<td>C7–T1</td>
<td>36, M</td>
<td>SAH, quadriplegia, BBD</td>
<td>PMV</td>
<td>cervical DAVF</td>
</tr>
<tr>
<td></td>
<td>C4–5</td>
<td>57, M</td>
<td>hemiparesis, BBD</td>
<td>epidural veins</td>
<td>epidural AVSs</td>
</tr>
<tr>
<td>Yamada et al., 1991</td>
<td>C5–6</td>
<td>46, F</td>
<td>quadriplegia</td>
<td>epidural veins, epidural AVSs</td>
<td>epidural AVSs</td>
</tr>
<tr>
<td>Morimoto et al., 1992</td>
<td>rt C-5</td>
<td>61, M</td>
<td>SAH</td>
<td>PMV, CVP</td>
<td>radicular AVM</td>
</tr>
<tr>
<td>Glasser et al., 1993</td>
<td>C4–5</td>
<td>9, M</td>
<td>suspected hemiparesis (thrombosis of draining vein, intramedullary hemorrhage)</td>
<td>PMV, venous aneurysm</td>
<td>perimedullary AVF</td>
</tr>
<tr>
<td>Kohno et al., 1996</td>
<td>rt C-3</td>
<td>51, M</td>
<td>radiculopathy, weakness of lt arm</td>
<td>epidural veins</td>
<td>epidural AVSs</td>
</tr>
</tbody>
</table>

* BBD = bowel-bladder disturbance; CVP = cervical venous plexus.
tion is related to mass effect rather than venous congestion. Therefore, one may argue that these cases may represent epidural AVSs rather than typical spinal DAVFs. In 1992, Morimoto et al. reported on a patient in whom angiography studies demonstrated a conglomerate of abnormal vessels surrounding the nerve root at the right C-5 level that were supplied by a radicular artery. Because of the nidal aspect of the vascular malformations, the characteristics of a radicular AVM are here fulfilled. One 9-year-old boy reported by Glasser et al. in 1993 had a high-flow AVS at the C4–5 level that was more characteristic of a perimedullary AVF. Only one of the patients described by Willinsky et al., who presented with SAH from rupture of a venous aneurysm before later developing quadriparesis related to congestive myelopathy, had an AVS at the right C7–T1 level, with the converging appearance of the radicular feeding vessels and slow-flow drainage of the veins, as typically encountered in spinal DAVF. Given these considerations, we presume that cervical spine DAVF is an exceedingly rare finding, which is further attested by the large series on spinal DAVF in which no cervical spinal DAVF was present in series of 49, 80, and 144 patients each. In our own experience of 40 patients with spinal DAVF seen over the past 10 years, this is the first one with a cervical lesion. Spinal DAVFs may occur anywhere from the level of the foramen magnum to the sacrum, and localization of these lesions may be difficult and challenging, as in our case, in which the myelopathy occurred distant to the AVS location. Advances in noninvasive, dynamic spinal MR angiography studies have greatly contributed to localizing these lesions, in helping to avoid unnecessary superselective injections of all possible arterial feeding vessels, and in directing the invasive and time-consuming spinal digital subtraction angiography.

In our case, the marked delay in the venous return after ASA injection indicated a diminished arteriovenous gradient due to arterialization of the veins, and this led us to the further investigation. While a delayed venous return warrants further searching for shunting spinal lesions, in cases with a normal venous phase after ASA injection, the diagnosis of a spinal DAVF can be excluded with a high degree of confidence.

Treatment of cervical DAVFs and cervical spine DAVFs is usually either surgical or endovascular. The goal of the treatment is to close the proximal vein, which may be done by surgical cauterization, clipping, or embolization accomplished using permanent liquid embolic materials such as N-butyl-cyanoacrylate or Onyx. However, if occlusion of the proximal vein is not achieved via an endovascular approach, fistulas will recruit collateral vessels from adjacent levels, and this will result in recurrent arteriovenous shunting. In our practice, in patients in whom the glue did not reach the proximal venous part, surgery is always performed to complete treatment of these lesions.

Conclusions

Lower cervical DAVFs are extremely rare and can be a challenging diagnosis when presenting with symptoms of lower-thoracic venous congestion. They have to be differentiated from other vascular lesions, such as radicular AVMs, epidural AVSs, and perimedullary AVFs. Surgery and endovascular treatment are both effective in treating these lesions, with the goal of both approaches being the complete occlusion of the shunting zone and the proximal part of the draining vein.

Disclaimer

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

References

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