Tentorial Dural Arteriovenous Fistula Presenting with Venous Congestive Edema of the Upper Cervical Cord

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Abstract

Tentorial dural arteriovenous fistula (DAVF) is an aggressive vascular lesion causing progressive neurological deficits. Venous congestive cervical edema is a rare phenomenon caused by tentorial DAVF. Obliteration of the fistula and venous drainage should be the goal of treatment. A 62-year-old man was admitted with lower limb weakness and numbness. Magnetic resonance imaging (MRI) revealed extensive edema of the upper cervical cord with signal flow void at the anterior spinal cord. Internal carotid angiography revealed a tentorial arteriovenous shunt near the superior petrosal sinus fed mainly by the tentorial artery. The petrosal vein was dilated, with the transverse pontine vein, medial medullary vein, and anterior spinal vein as the main drainage route. This suggests that venous hypertension triggered the upper cervical cord edema. MRI with gadolinium enhancement showed that the varix was located just distal to the shunt. Microsurgical obliteration of the fistula and venous drainage were achieved via a suboccipital approach. A postoperative evaluation showed the disappearance of the cervical cord edema. MRI with gadolinium enhancement showed that the varix was located just distal to the shunt. Microsurgical obliteration of the fistula and venous drainage were achieved via a suboccipital approach. A postoperative evaluation showed the disappearance of the cervical cord edema with improved clinical symptoms. Tentorial DAVF with spinal venous drainage presents with mild and slow progression of symptoms. Differential diagnosis and definite treatment are mandatory to avoid a delayed diagnosis and irreversible symptoms.

Keywords: arteriovenous fistula, tentorial artery, venous congestion, upper cervical

Introduction

Tentorial dural arteriovenous fistula (DAVF) has angiographic features of retrograde drainage through cortical or subarachnoid veins, deep drainage through the vein of Galen, and venous varices. It is characterized by a variety of symptoms, including headache, dizziness, nausea, vomiting, gait instability, pulsatile tinnitus, dysarthria, and myelopathy. Spinal venous drainage in tentorial DAVF contributes to myelopathy, but its severity and progression depend on the venous hypertension and cord congestion. Mild symptoms cause a delay in the clinical diagnosis and irreversible symptoms.

We herein report a case of tentorial DAVF with edema of the upper cervical cord due to venous drainage into the spinal veins with gradually apparent myelopathy after more than one year.

Case Report

A 62-year-old man had noticed bilateral foot numbness 2 years earlier. He had visited an orthopedic hospital. Magnetic resonance imaging (MRI) showed mild canal stenosis. His symptom was diagnosed as lumbar canal stenosis. Medication was administered and physical therapy was performed, but his symptoms gradually developed. He underwent lumbar decompression surgery, but his symptoms progressed upward to the calves over time. On referral to our hospital, he presented with bilateral numbness up to the thigh, bilateral hand coldness, and positive Romberg sign. His bilateral patella tendon reflex increased. Urinary disturbance was also noted. Muscle weakness in the upper and lower extremities was not present. Routine laboratory examinations yielded normal results. The patient was non-diabetic and non-rheumatoid. Neurological examinations indicated myelopathy due to a cervical cord lesion. He had
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Fig. 1  A: Preoperative sagittal T2-weighted magnetic resonance imaging revealing swelling and an abnormal intensity of the upper cervical spinal cord. B: Lateral view of preoperative right internal carotid angiography showing the varix (asterisk). The feeding system was composed of the tentorial artery (arrowhead) and the feeder from the meningohypophyseal trunk (double arrowheads). C: Anterior-posterior view of preoperative right internal carotid angiography showing TPV (double arrows) as the main draining route into the AMPV. D: Preoperative gadolinium-enhanced CT revealing the varix (asterisk) and petrosal vein (drainer; arrow) around the right trigeminal nerve. Each arrow and arrowhead in A-E show the same vascular structures. E: A schematic illustration of the preoperative angiographic interpretation.

The patient’s neurological symptoms gradually improved after the surgery, except for his urinary disturbance. Postoperative MRI revealed the resolution of the T2-hyperintense signal, swelling of the spinal cord, and abnormal dilated spinal vein disappearance (Fig. 3A). Follow-up angiography performed 1 month after the surgery confirmed DAVF disappearance (Fig. 3B, C). The feeding artery from the petrosal branch of the middle meningeal artery disappeared. The patient was transferred to a rehabilitation division 1 month after the surgery.

This study was approved by the ethical review board of Osaka Gyoumeikan Hospital. The patient provided informed consent prior to inclusion in this study.

Discussion

The present case had upper cervical cord edema with a flow void signal of the anterior spinal veins at the C3/4 spinal level. These MRI findings suggested the existence of vascular abnormalities of the upper cervical spine. However, we found a tentorial DAVF draining into the anterior spinal vein. Furthermore, the onset symptom was bilateral foot numbness and myelopathy that gradually appeared over one year. This is a rare case of tentorial DAVF with
Tentorial DAVF with Cervical Cord Edema

The present patient had a tentorial DAVF with edema of the upper cervical cord due to venous drainage into the spinal veins, which gradually manifested as cervical myelopathy over one year. In our extensive research on PubMed, among tentorial DAVFs, a total of 14 surgical cases have been associated with cervical congestion due to a spinal draining vein (Table 1). The arterial supply of a tentorial DAVF is extensive, involving meningeal arteries from the internal carotid artery and vertebral artery that are riskier to embolize than external carotid artery feeders. Transvenous navigation to deeper locations around the tentorium is difficult. Furthermore, subarachnoid veins in tentorial DAVF prevent transvenous access. Therefore, embolization management was only feasible in three cases of tentorial DAVF with venous drainage into spinal veins (Table 1). In the present case, the draining route involved petrosal, perimedullary, and spinal veins. The draining petrosal veins were relatively small. Arterial embolization carries a risk of neurological complications and difficulty with cannulation. We therefore performed open surgery for the complete obliteration of the fistula and venous drainage.

Tentorial DAVF results in the most aggressive neurological behavior, with 97% causing hemorrhaging or progressive focal neurological deficits. However, in cases of tentorial DAVF with venous drainage into the spinal veins, the progression of symptoms was slow, with various symptoms showing an onset. Eight of the 14 cases took several months from the onset of symptoms to achieve a diagnosis of tentorial DAVF (Table 1). These cases presented with and gradually developed numbness, incoordination, hiccup, and dizziness. Tentorial DAVF with venous drainage into the spinal veins has no hemorrhaging. In the present case, the neurological symptom at the onset was bilateral foot numbness. The prompt diagnosis of these arteriovenous shunts is sometimes difficult, as medical conditions that can cause foot numbness include peripheral vascular disease, diabetic neuropathy, lumbar canal stenosis, herniated disc, multiple sclerosis, vasculitis, myelitis, and side effects of chemotherapy medication.

It is very difficult to promptly diagnose tentorial DAVF causing cervical spinal cord edema at the very beginning of clinical practice. The lateral mesencephalic vein (LMV) represents a connection between the ventral diencephalic segment of the basal vein and ventral mesencephalic veins. The LMV drains in three main directions. The LMV runs upward and drains into the basal vein or posterior mesencephalic vein, drains inferiorly into the petrosal vein, and

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Fig. 2 A: Intraoperative photograph showing the varix (asterisk) and petrosal vein (arrow) over the trigeminal nerve. B: Operative field illustration. C: Intraoperative indocyanine green video angiography showing the drainer running down from the varix to the petrosal vein. D: Final view of the operation showing the coagulated varix and disconnected petrosal vein.
Fig. 3  A: Sagittal T2-weighted magnetic resonance imaging obtained 1 month after treatment showing the marked resolution of the upper cervical spinal cord swelling and edema. B, C: Follow-up right internal carotid angiography performed 1 month after treatment showing no residual shunt.

Table 1  Clinical summary of 14 patients with tentorial DAVF with spinal venous drainage

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No.</th>
<th>Age (Yrs)</th>
<th>Sex</th>
<th>Feeders</th>
<th>Drainers</th>
<th>Treatment</th>
<th>Time to diagnosis*</th>
<th>Onset symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wrobel et al., 1988</td>
<td>1</td>
<td>43</td>
<td>M</td>
<td>OA, TA</td>
<td>PV → SV</td>
<td>E and S</td>
<td>2 years</td>
<td>Leg incoordination</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>68</td>
<td>M</td>
<td>OA, TA</td>
<td>PV → PMV → SV</td>
<td>S</td>
<td>6 months</td>
<td>Foot coldness, thigh numbness</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>42</td>
<td>M</td>
<td>OA, PA</td>
<td>SV</td>
<td>S</td>
<td>1 month</td>
<td>Urinary disturbance</td>
</tr>
<tr>
<td>Bret et al., 1994</td>
<td>4</td>
<td>31</td>
<td>M</td>
<td>MHT</td>
<td>TS, SV</td>
<td>S</td>
<td>4 months</td>
<td>Gait disturbance</td>
</tr>
<tr>
<td>Bousson et al., 1999</td>
<td>5</td>
<td>36</td>
<td>M</td>
<td>multiple</td>
<td>SV</td>
<td>S</td>
<td>1 year</td>
<td>Leg and arm numbness</td>
</tr>
<tr>
<td>Wiesmann et al., 2000</td>
<td>6</td>
<td>46</td>
<td>M</td>
<td>APA</td>
<td>PMV → SV</td>
<td>E</td>
<td>5 days</td>
<td>Headache, urinary disturbance</td>
</tr>
<tr>
<td>Pannu et al., 2004</td>
<td>7</td>
<td>42</td>
<td>M</td>
<td>TA</td>
<td>SV</td>
<td>S</td>
<td>1 year</td>
<td>Incoordination, dizziness</td>
</tr>
<tr>
<td>Khan et al., 2009</td>
<td>8</td>
<td>20</td>
<td>F</td>
<td>TA</td>
<td>CV, SV</td>
<td>S</td>
<td>4 weeks</td>
<td>Leg pain and tingling</td>
</tr>
<tr>
<td>Yamaguchi et al., 2009</td>
<td>9</td>
<td>79</td>
<td>M</td>
<td>TA, MMA, SCA</td>
<td>PMV → SV</td>
<td>S</td>
<td>3 months</td>
<td>Hand shaking</td>
</tr>
<tr>
<td>Takeshita et al., 2011</td>
<td>10</td>
<td>68</td>
<td>M</td>
<td>MMA, MHT, AMA</td>
<td>SV</td>
<td>E and S</td>
<td>1 year</td>
<td>Hiccup</td>
</tr>
<tr>
<td>Gross et al., 2014</td>
<td>11</td>
<td>69</td>
<td>M</td>
<td>TA, MMA, OA</td>
<td>SV</td>
<td>E</td>
<td>N.D.</td>
<td>Hand and arm pain</td>
</tr>
<tr>
<td></td>
<td>12</td>
<td>34</td>
<td>F</td>
<td>OA</td>
<td>SPS, PV → PMV → SV</td>
<td>E</td>
<td>N.D.</td>
<td>Leg and arm weakness</td>
</tr>
<tr>
<td>Rubio et al., 2019</td>
<td>13</td>
<td>68</td>
<td>M</td>
<td>MMA</td>
<td>PMV → SV</td>
<td>E and S</td>
<td>N.D.</td>
<td>Leg weakness</td>
</tr>
<tr>
<td>Present case</td>
<td>14</td>
<td>62</td>
<td>M</td>
<td>TA, MMA, MHT</td>
<td>PV → PMV → SV</td>
<td>S</td>
<td>1.5 years</td>
<td>Foot numbness</td>
</tr>
</tbody>
</table>

* Time from onset of symptoms to diagnosis; AMA = accessory meningeal artery; CV = cerebellar vein; E = embolization; PA = pharyngeal artery; PMV = perimedullary vein; PV = petrosal vein; S = surgery; SCA = superior cerebellar artery; SPS = superior petrosal sinus; SV = Spinal vein; TS = Transverse sinus
drains directly in the petrosal vein when the basal vein is hypoplastic or absent. The LMV is involved in the venous drainage of 31% of patients with tentorial DAVFs. Table 1 shows that the perimecencephalic vein is involved in the venous drainage of 42.8% of tentorial DAVFs with spinal venous drainage, whereas the petrosal vein is involved in the venous drainage of 28.5%. A venous varix and vascular dilated anomaly of the pontomesencephalic junction in intracranial brain MRI may help to elucidate early diagnosis for tentorial DAVF with spinal venous drainage.

Spinal DAVF is a slow-flow extramedullary vascular lesion typically affecting the lower thoracic and lumbar spinal levels. Spinal DAVF does not usually occur with hemorrhaging. Progression to severe myelopathy is slow. Patients with spinal DAVF initially present with lower-extremity dysesthesias and intermittent radicular pain mimicking peripheral nerve lesions. These vague symptoms make the diagnosis complicated and sometimes delay it. Many of these patients have already suffered from their symptoms for several months and have undergone unnecessary procedures and surgery before a diagnosis is finally made. The onset and progression of symptoms in spinal DAVF is similar to those in tentorial DAVF with spinal drainage.

Tentorial DAVF manifests with hemorrhaging or progressive focal neurological deficits. However, tentorial DAVF with spinal venous drainage occasionally presents with mild and slow progression of symptoms. Tentorial DAVF may be a differential diagnosis of spinal DAVF.

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Abbreviations

DAVF; dural arteriovenous fistula, MRI; magnetic resonance imaging, ICG; indocyanine green

Conflicts of Interest Disclosure

No other disclosures reported.

References


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