Preoperative Evaluation and Surgical Strategies for Craniocervical Junction Dural Arteriovenous Fistula with Multiple Feeders: Case Report and Review of the Literature

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Case Report

Preoperative Evaluation and Surgical Strategies for Craniocervical Junction Dural Arteriovenous Fistula with Multiple Feeders: Case Report and Review of the Literature

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Abstract

Craniocervical junction dural arteriovenous fistula (CCJDAVF) fed by bilateral vertebral arteries (VAs) is extremely rare. We report a case of a 63-year-old man presenting with progressive myelopathy caused by a CCJDAVF, which was fed by bilateral VAs and occipital and ascending pharyngeal arteries with multiple shunting points and that drained into intracranial sinus and spinal veins. The dural arteriovenous fistula (DAVF) was successfully treated surgically using stepwise indocyanine green (ICG) videoangiography. After surgery, the DAVF disappeared and myelopathy was markedly improved. We show detailed preoperative images and intraoperative findings of this rare DAVF and emphasize the importance of selective angiography for preoperative evaluation of feeding arteries and the usefulness of intraoperative ICG videoangiography for both identification of the fistula and confirmation of its obliteration.

Key words: Dural arteriovenous fistula ・ Craniocervical junction ・ ICG videoangiography ・ Varix, Vertebral artery

Dural arteriovenous fistula (DAVF) is the most frequent spinal vascular malformation. DAVF is located in the dura mater around the nerve root with communication between a radiculomeningeal artery and a radicular vein that drains into perimedullary veins. DAVF usually occurs in the thoracolumbar region and less commonly at the craniocervical junction. Generally, craniocervical junction DAVF (CCJDAVF) is supplied by one or two meningeal branches of the unilateral vertebral artery (VA) and occasionally receives an additional supply from the ipsilateral occipital or ascending pharyngeal arteries. Only three cases of CCJDAVF fed by bilateral meningeal branches of the VA have been reported. We report here an extremely rare case of CCJDAVF fed by bilateral posterior meningeal arteries arising from VAs and a meningeal branch arising from occipital and ascending pharyngeal arteries.

Preoperative evaluation of the vascular structure and the surgical strategy using ICG videoangiography are described in detail.

Case report

History and examination

A 63-year-old man presented for the first time with numbness in the right lower extremity. Three months later, the numbness had extended to the left lower extremity. Ten months later, he presented with progressive intermittent claudication and was admitted to the hospital. He had no history of head trauma or cerebrovascular disease. Neurological examination revealed se-
Fig. 1  (A) Preoperative T2–weighted sagittal image showing marked swelling of the spinal cord with edema above T2 extending to the medulla oblongata. (B) Gadolinium–enhanced T1–weighted sagittal image showing enlarged and tortuous vessels along the surface of the cervical spinal cord and a varix (white arrow) at the dorsal surface of the medulla oblongata. (C) MR cisternography showing that the varix (black arrow) was connected to the dura mater of the posterior fossa.

Fig. 2  Preoperative digital subtraction angiography showing anteroposterior (AP) view of right (A) and left (B) vertebral angiography (VAG), lateral view of left VAG (C), AP view of left external carotid angiography (D), AP view of right (E) and left (F) posterior meningeal artery angiography, and lateral view of left posterior meningeal angiography (G), demonstrating a dural arteriovenous fistula (DAVF) fed by bilateral posterior meningeal arteries (arrowhead) of the vertebral arteries and a meningeal branch (double arrowheads) formed by the left occipital and ascending pharyngeal arteries, draining into the right transverse sinus via an occipital sinus (arrow), left transverse sinus (double arrows) via the prepontine venous system (asterisk), and anterior and posterior spinal veins (double asterisks) via perimedullary veins.
vere hypoesthesia and continuous numbness below the L2 dermatome as well as manual muscle testing (MMT) 4/5 muscle strength and enhanced spasticity in the lower extremities with positive Babinski sign. No bladder or rectal functional disruptions and no lower cranial nerve dysfunction were observed. T2-weighted magnetic resonance (MR) imaging demonstrated swelling with high signal intensity accompanied by dotted low signal intensities in the cervical spinal cord and medulla oblongata (Fig. 1A). Gadolinium-enhanced T1-weighted MR imaging demonstrated enlarged and tortuous vessels along the surface of the cervical spinal cord and a varix at the dorsal surface of the medulla oblongata (Fig. 1B). MR cisternography revealed that the varix arose from the dura mater of the posterior fossa (Fig. 1C). Vertebral digital subtraction angiography (DSA) revealed a CCJDAVF, which was fed by bilateral posterior meningeal arteries from VAs at the same vertebral level and that drained into the dilated anterior and posterior spinal veins with formation of a varix (Fig. 2A–C). Furthermore, selective left external carotid angiography revealed another feeding artery formed by the union of a meningeal branch of the left occipital artery and a neuromeningeal branch of the left ascending pharyngeal artery (Fig. 2D). Selective DSA at the origin of the meningeal arteries from the VAs revealed that the DAVF was also draining into the right transverse sinus via an occipital sinus and left transverse sinus via a perimesencephalic vein through the preoptic venous system (Fig. 2E–G). For more detailed evaluation, image fusion of 3D-DSA and 3D-computed tomography (CT) angiography was performed with workstation software (Zio Station; Zio Soft, Tokyo, Japan) (Fig. 3A, B). 3D-reconstructed fusion images revealed that the lesion was located in the dura mater around the midline of the craniocervical junction with multiple shunt points.

**Operation and postoperative course**

The patient was placed in the prone position. A midline suboccipital craniotomy and C1 laminotomy were performed. The origin of the bilateral posterior meningeal arteries was exposed (Fig. 4A). Before opening the dura, ICG videoangiography was performed, and the entire fistula and the three feeding arteries that ran within the dura mater into the fistula were clearly identified from

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**Fig. 3** Dorsal view (A) and intracranial view (B) of reconstructed fusion images of 3D-DSA and 3D-CT angiography revealing that the DAVF was located at the dura mater around the midline of the craniocervical junction with multiple shunt points. The arrowhead shows the bilateral posterior meningeal arteries from the vertebral arteries. The double arrowheads show a meningeal branch of the occipital artery (arrow) and the neuromeningeal branch of the ascending pharyngeal artery (double arrows), passing through the mastoid foramen (asterisk) and jugular foramen (double asterisks), respectively.
the dorsal surface of the craniocervical junction (Fig. 4B). Then, the dura mater was partially incised, and the three main feeding arteries were temporarily occluded with clips that were applied through the cut edge of the dura mater (Fig. 4C). Subsequent ICG videoangiography demonstrated subtle enhancement of the fistula, draining vein, and a varix in the late venous phase (Fig. 4D). Then, all feeding arteries were permanently obliterated, and the dura mater was sufficiently coagulated around the fistula. After the dura mater was incised to surround the fistula, a draining vein and the varix were exposed (Fig.
Then, the draining vein was disconnected at its origin using an aneurysmal clip (Fig. 5B). ICG videoangiography demonstrated disappearance of the blood flow draining from the varix (Fig. 5C). The postoperative course was uneventful, and the symptoms before the operation immediately improved. Postoperative DSA revealed complete disappearance of the DAVF (Fig. 6A–C). T2-weighted MR images also demonstrated reduction of intramedullary high intensity and disappearance of flow voids around the spinal cord (Fig. 6D).

**Discussion**

AVF at the craniocervical junction is uncommon. To our knowledge, only 79 cases including ours have been reported.1–12 Table 1 summarizes the cases of DAVF at the craniocervical junction in the literature, listing age, sex, vascular supply and drainage route, and the presence of a varix and subarachnoid hemorrhage (SAH). Excluding two cases in which the gender was unspecified, the 77 patients included 57 men (72.2%) and 20 women (15.2%). The age of the patients ranged from 30 to 85 years with an average of 57.3 years (57 years for men, 58.2 years for women). Thirty-one patients (39.2%) presented with SAH. Excluding cases in which the presence of a varix was unspecified (15 of the 79 cases), a varix was found in 18 (28.1%) of 64 patients. In 18 patients with a varix, 14 patients (77.8%) presented with SAH. On the other hand, in 46 patients without a varix, 12 patients (26.1%) presented with SAH. The presence of a varix is considered to be strongly associated with presence of SAH.

Venous drainage of the CCJDAVF is via an ascending drainage route into the intracranial sinus and/or a descending drainage route into the spinal medullary vein.13–20 An ascending drainage route is considered to be associated with varix formation and presence of SAH, whereas a descending drainage route is considered to be associated with presence of myelopathy.13–20 Of the 70 previous cases in the literature in which the drainage route was specified, venous drainage was via an ascending route in 29, both ascending and descending routes in 11, and a descending route in 30. Twenty of 29 patients (69%) with an ascending drainage route, four of 11 patients (36.4%) with ascending and descending routes, and five of 30 patients (16.7%) with a descending route presented with SAH. In our case, the main drainage route was spinal veins, and the patient had presented with progressive myelopathy. However, the patient also had a high risk of SAH because he had a varix and an ascending drainage route into the intracranial sinus.

The arterial supply was assessed in 76 cases in the literature. Approximately 95% of CCJDAVF cases were fed by a branch of the VA including the C1 or C2 radicular artery, the meningeal or muscular branch, and the anterior or posterior meningeal artery. The fistula was fed by a unilateral VA in most cases. However, a fistula fed by bilateral VAs was seen in four cases including ours. Other arterial supplies were the ascending pharyngeal artery in 23 cases (30.3%), occipital artery in 12 cases (15.8%), middle meningeal artery in three cases, meningo-hypophyseal trunk in three cases, posterior inferior cerebellar artery in two cases, and posterior auricular artery in two cases. CCJDAVF with multiple feeding arteries was seen in 19 cases (25%). Among them, CCJDAVF with three or more feeders was seen in eight cases including ours (10.5%). Therefore, if a CCJDAVF was identified by a unilateral vertebral angiogram, another arterial supply should be carefully evaluated by selective injection of the contralateral VA and bilateral internal and external carotid arteries.

In this case, ICG videoangiography was used for both identification of the fistula and vascular structures and confirmation of its obliteration during surgery. Intraoperative DSA requires preoperative placement of a catheter within the feeding artery. Therefore, careful preoperative planning and technical expertise are required, especially when the patient is in the prone position.
Table 1  Summary of the reported cases of dural arteriovenous fistula at the craniovertebral junction

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Feeding arteries</th>
<th>Draining veins</th>
<th>Drainage varix</th>
<th>SAH</th>
</tr>
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<tr>
<td>Asakawa et al., 2002</td>
<td>64, M</td>
<td>APA</td>
<td>MV</td>
<td>D</td>
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<td>Aviv et al., 2004</td>
<td>57, M</td>
<td>VA meningal br</td>
<td>Epidural vein</td>
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<td>no</td>
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<td>Barnwell et al., 1990</td>
<td>69, M</td>
<td>VA br, APA</td>
<td>MV</td>
<td>US</td>
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<td>yes</td>
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<tr>
<td>Chen et al., 1998</td>
<td>36, M</td>
<td>MMA, VA meningal br</td>
<td>IVV, ant. SV</td>
<td>A/D</td>
<td>no</td>
<td>no</td>
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<tr>
<td>Choi et al., 2012</td>
<td>47, M</td>
<td>VA br</td>
<td>IVV, ant. &amp; post. SV</td>
<td>A/D</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>Do et al., 1999</td>
<td>50, M</td>
<td>VA muscular br</td>
<td>ant. MV, ICV</td>
<td>A</td>
<td>no</td>
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<td>Ernst et al., 1997</td>
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<tr>
<td>Fox et al., 1978</td>
<td>54, M</td>
<td>APA</td>
<td>IJV</td>
<td>D</td>
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<td>VA br, APA</td>
<td>MV</td>
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<tr>
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<td>IJV</td>
<td>D</td>
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<tr>
<td>Hahn et al., 1998</td>
<td>67, M</td>
<td>APA, OA</td>
<td>SPS, ant. &amp; post. SV</td>
<td>A/D</td>
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<td>no</td>
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<tr>
<td>Hanabusa et al., 1999</td>
<td>61, M</td>
<td>VA meningal br</td>
<td>ICV</td>
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<td>C1 RA</td>
<td>ICSV, perimedullary vein</td>
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<td>Javier et al., 1999</td>
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<td>US</td>
<td>MV</td>
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<td>VA br</td>
<td>ant. &amp; post. SV</td>
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<tr>
<td>Oda et al., 1989</td>
<td>58, M</td>
<td>VA br</td>
<td>CVP, Epidural vein</td>
<td>A</td>
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<td>Ogawa et al., 2012</td>
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<td>C1 RA</td>
<td>CVP, MV</td>
<td>D</td>
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<tr>
<td>Niwa et al., 1997</td>
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<td>MV</td>
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<td>Nonaka et al., 1999</td>
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<td>C1–C2 RA</td>
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position. Moreover, in cases with multiple feeding arteries such as ours, it is difficult to perform intraoperative selective injection of all feeding arteries. In our case, using ICG videoangiography, we could easily identify the location, extent, and blood supply of the DAVF before dural incision (Fig. 4B). ICG videoangiography through the dura mater allowed us to plan an adequate dural incision around the fistula and to coagulate the fistula completely.

**Conclusion**

We demonstrated a rare case of CCJDAVF with multiple feeders including bilateral VAs, the ascending pharyngeal artery, and the occipital artery. In the preoperative angiographic evaluation for CCJDAVF, selective injection of the bilateral internal carotid artery, external carotid artery, and VA is necessary for detection of the feeding arteries. Intraoperatively, repetitive ICG videoangiography is quite useful for understanding the structure, flow dynamics, and confirmation of complete obliteration of DAVF.

**Conflicts of interest : None**

**Acknowledgments : None**
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両側の椎骨動脈を栄養動脈とする頭蓋頸椎移行部硬膜動静脈瘻（CCJDAVF）は極めて稀な病態である。本症例は63歳男性で、進行性脊髄症により見つかったCCJDAVFである。両側椎骨動脈をはじめとした複数の栄養動脈を有し、複数のシャントポイントを介して頭蓋内静脈洞および脊髄静脈へと流出する稀な病態を呈しており、直達手術により完治した。手術では、術中インドシアングリーン（ICG）蛻光血管撮影を繰り返し用いて、CCJDAVFの解剖学的構造および血流動態を把握しながら段階的に栄養動脈を処理し、最終的に流出静脈を遮断した。最後に再びICG蛻光血管撮影を行い、CCJDAVFの消失を確認した。術後、脊髄症は著明に改善した。複数の流入動脈およびシャントポイントを有する本症例の複雑な病態把握においては、術前の選択的血管撮影を用いた流入動脈の評価が重要であった。術中ICG蛻光血管撮影を繰り返し用いることにより、病変の同定および基孔の消失を確実に行うことが可能であった。