Dural arteriovenous fistula of the anterior condylar confluence and hypoglossal canal mimicking a jugular foramen tumor

Case report

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The anterior condylar confluence (ACC) is located on the external orifice of the canal of the hypoglossal nerve and provides multiple connections with the dural venous sinuses of the posterior fossa, internal jugular vein, and the vertebral venous plexus. Dural arteriovenous fistulas (DAVFs) of the ACC and hypoglossal canal (anterior condylar vein) are extremely rare. The authors present a case involving an ACC DAVF and hypoglossal canal that mimicked a hypervascular jugular bulb tumor.

This 53-year-old man presented with right hypoglossal nerve palsy. A right pulsatile tinnitus had resolved several months previously. Magnetic resonance imaging demonstrated an enhancing right-sided jugular foramen lesion involving the hypoglossal canal. Cerebral angiography revealed a hypervascular lesion at the jugular bulb, with early venous drainage into the extracranial vertebral venous plexus. This was thought to represent either a glomus jugulare tumor or a DAVF.

The patient underwent preoperative transarterial embolization followed by surgical exploration via a far-lateral transcandylar approach. At surgery, a DAVF was identified draining into the ACC and hypoglossal canal. The fistula was surgically obliterated, and this was confirmed on postoperative angiography. The patient’s hypoglossal nerve palsy resolved.

Dural arteriovenous fistulas of the ACC and hypoglossal canal are rare lesions that can present with isolated hypoglossal nerve palsies. They should be included in the differential diagnosis of hypervascular jugular bulb lesions.

The authors review the anatomy of the ACC and discuss the literature on DAVFs involving the hypoglossal canal. (DOI: 10.3171/JNS/2008/109/8/0335)

Key Words • anterior condylar confluence • dural arteriovenous fistula • hypoglossal canal • jugular foramen tumor

D u ral arteriovenous fistulas are aberrant vascular connections shunting arterial inflow directly to venous outflow and are usually confined within a dural venous sinus. Symptoms of DAVFs typically correlate with their pattern of venous drainage. The lesions commonly occur in the cavernous sinus, presenting with ocular symptoms, and also in the sigmoid and transverse sinuses, presenting most frequently with a pulse-synchronous bruit. Dural arteriovenous fistulas that involve the transverse or sigmoid sinuses comprise the majority that occur in the posterior fossa, but there are less common fistulas that occur at the skull base, including DAVFs of the marginal sinus at the foramen magnum.5–9 These malformations can also develop wherever veins follow a transosseous pathway.7 Such is the case with DAVFs that drain into the anterior condylar vein, which courses within the osseous hypoglossal canal.2,3,5,11 These fistulas involving the hypoglossal canal (anterior condylar vein) are extremely rare; only several cases have been reported.2,3,5,11 We report the case of a patient who presented with an isolated hypoglossal nerve palsy and in whom a DAVF was found to drain into the ACC and through the hypoglossal canal. This case is unique in that the DAVF mimicked a hypervascular jugular bulb tumor and was treated with a combined endovascular and skull base approach to obliterate the fistula.

Case Report

History and Examination. This 53-year-old man presented with a several-year history of right-sided pulsatile tinnitus that had resolved on its own. After resolution of the tinnitus, he noticed some tongue weakness and dysarthria while singing and speaking. On examination, he
exhibited tongue atrophy on the right half and deviation of the tongue to the right, consistent with a right-sided hypoglossal nerve palsy. An audible bruit was present behind the right ear. The remaining neurological examination findings were normal.

Brain MR imaging demonstrated a 1.9 × 2.5-cm lesion at the jugular foramen involving the right hypoglossal nerve canal; we observed an intermediate T1 signal with enhancement, suspicious for a highly vascular lesion (Fig. 1). The radiological differential diagnosis included metastatic tumor, glomus tumor, and schwannoma. Computed tomography of the chest, abdomen, and pelvis was performed to rule out metastatic disease, and no abnormal lesions were found.

Cerebral angiography demonstrated a highly vascular lesion at the right jugular bulb. The main arterial supply was from the right ascending pharyngeal artery, and blood was additionally supplied by the right posterior auricular and the occipital arteries (Figs. 2 and 3). There was early shunting of blood from the feeding arteries directly into the venous system primarily via markedly abnormal extracranial veins, suggestive of a high-flow lesion. We believed that this represented a hypervascular lesion at the jugular bulb consistent with a high-flow glomus tumor. The lesion did not seem to have the features of a DAVF because of its high-flow nature, although this entity was included in the differential diagnosis.

Treatment. The patient underwent preoperative transarterial embolization of the 3 main arterial feeding structures to decrease the blood supply to the presumed tumor. There was minimal residual filling of the lesion after embolization.

Operation. The patient underwent a right-sided far-lateral transcylindrical approach, including a partial mastoidectomy, removal of the posterior one-third of the occipital condyle, and reduction of the jugular tubercle for exposure of the jugular bulb and hypoglossal canal (Figs. 4 and 5). The mastoid segment of the facial nerve was skeletonized down to the region of the jugular bulb. In this region, a vascular structure located in the leaflets of the dura mater was identified. This structure was incised and still had an active vascular supply, which was controlled with bipolar cautery. A large draining vein was followed from this vascular structure in the jugular bulb distally down to the venous plexus in the hypoglossal canal. It was anatomically consistent with the ACC draining from the jugular bulb and continuing as a dilated anterior condylar vein within the hypoglossal canal. The fistula was obliterated by occluding and dividing the venous outflow at the ACC (Fig. 5).

Postoperative Course. The patient did well after surgery. The right hypoglossal nerve palsy resolved, and there was no cerebrospinal fluid leak. The remainder of the hospital course was uneventful and the patient was discharged home on the 3rd postoperative day. Postoperative angiography showed complete obliteration of the DAVF (Fig. 6).

Discussion

Review of the Literature

Dural arteriovenous fistulas of the anterior condylar vein and hypoglossal canal are a rare subtype of posterior fossa DAVF; 8 cases (including ours) have been reported.2,3,5,10 In a review of these 8 cases (Table 1), we determined a mean age at presentation of 65 years (range 43–83 years) and no sex predominance (4 men and 4 women). Clinical presentation was highly variable and appeared to reflect the pattern of venous drainage. Seven patients (88%) presented with a pulse-synchronous bruit or pulsatile tinnitus. Three patients (38%) presented with oculomotor symptoms similar to carotid–cavernous fistulas including chemosis, proptosis, and diplopia due to retrograde venous flow into the inferior petrosal sinus, cavernous sinus, and superior ophthalmic vein.3,5,10 One patient presented with cervical myelopathy due to spinal cord compression caused by a tortuous anterior spinal draining vein. Two patients (including ours) presented with an isolated hypoglossal nerve palsy. This was attributed to increased venous pressure from predominant drainage through a dilated anterior condylar vein within the hypoglossal canal. In both cases, the hypoglossal nerve palsy improved after obliteration of the fistula.

Dural arteriovenous fistulas can change over time, progressing from low- to high-risk lesions.7 This progression can manifest clinically as a loss or reduction in a bruit or tinnitus. When antegrade drainage into the jugular bulb and vein is obstructed or restricted, alternative venous outflow pathways are formed and venous hypertension can occur within these pathways. In the present

Fig. 1. Axial T1-weighted MR images demonstrating an enhancing lesion in the right jugular foramen (arrows), suggestive of a hypervascular jugular foramen tumor.
case, hypoglossal nerve palsy presented shortly after the patient’s pulsatile tinnitus abated. Angiographically, there was no flow into the jugular vein, but there was high-flow blood draining into a dilated anterior condylar vein causing hypoglossal nerve palsy.

**Anatomy of the ACC and Hypoglossal Canal**

Knowledge of the venous anatomy at the cranio cervical junction is key to understanding DAVFs of the ACC and hypoglossal canal. In 1868, Trolard described a venous confluence (an ACC) that was located extracranially in front of the aperture of the hypoglossal canal. The following veins contributed in forming the ACC: 1) the anterior condylar vein; 2) one or several branches from the internal jugular vein or bulb; 3) the lateral condylar vein, sometimes arising from one or several branches from the internal jugular vein or bulb; 4) anastomoses from the inferior petrosal sinus; 5) branches from the internal carotid artery venous plexus of Rektorzik; and 6) branches from the prevertebral venous plexus found on the anterior atlantoccipital membrane.

The ACC appears to be an anatomical constant whose major tributaries are the anterior and lateral condylar veins, inferior petrosal sinus, and internal jugular vein. It provides important connections between the intracranial cerebral venous circulation and the vertebral venous systems at the craniocervical junction. Venous outflow from the jugular bulb through the anterior condylar vein and into the vertebral venous plexus occurs by way of the ACC. This venous confluence becomes more clinically significant in pathological conditions such as DAVFs of the hypoglossal canal. In our case, the hypervascular blush represented the DAVF of the ACC with antegrade filling of the anterior condylar vein and extracranial vertebral venous plexus. The hypervascular blush at the ACC angiographically mimicked a high-flow jugulare glomus tumor. Interestingly, the venous drainage occurred via the anterior condylar vein and extracranial vertebral venous plexus, not through the jugular bulb and vein.

The hypoglossal canal is located inferomedial to the jugular foramen and is the conduit for the hypoglossal nerve, which is surrounded by the hypoglossal venous plexus, also referred to as the anterior condylar vein. This vein originates from the ACC near the junction of the jugular bulb and the inferior petrosal sinus and extends into the hypoglossal canal. It can communicate with the suboccipital venous plexus, vertebral or paravertebral venous plexus, posterior condylar vein, marginal sinus, basilar venous plexus, or the internal vertebral venous plexus.

Because of the numerous potential communications with surrounding venous structures, DAVFs of the anterior condylar vein can have various venous drainage patterns,
as was demonstrated in the 8 cases in the literature. The hypoglossal canal can also contain meningeal branches of the ascending pharyngeal artery that become clinically significant when they become feeding arteries of a DAVF.

Treatment of Hypoglossal Canal DAVFs

Several treatment options can be used to obliterate these DAVFs, including endovascular therapy (transvenous or transarterial embolization), surgery, or both.
The pattern of venous drainage can help determine the optimal treatment strategy. Transfemoral transvenous embolization is very effective and appears to be the treatment of choice in eliminating DA VFs; however, it is not always feasible if communication with the fistulous point via the jugular vein or bulb is absent. Alternatively, transarterial embolization can be performed, although this is generally considered less effective, with higher recurrence rates due to an inability to embolize the exact site of the fistula and all its feeding artery pedicles. Transarterial embolization, however, can be useful in reducing flow through the fistula before attempting definitive treatment. Direct surgical obliteration of DA VFs may be indicated to interrupt the venous outflow if endovascular intervention cannot eliminate the fistula. Stereotactic radiosurgery can be considered in cases in which the lesions are difficult to access surgically or via endovascular embolization. In the present case, transvenous embolization was not possible, and transarterial embolization resulted in partial treatment of the DAVF. The fistula was readily accessible and obliterated via a far-lateral transcondylar approach, providing exposure of the jugular bulb, anterior condylar vein, and hypoglossal canal.

**Conclusions**

Dural arteriovenous fistulas involving the ACC and hypoglossal canal are rare vascular lesions at the skull base that can present with an isolated hypoglossal nerve palsy. Their clinical manifestation appears to arise from compressive symptoms as a result of the venous drainage pattern. Successful treatment can be achieved with transvenous or transarterial embolization, surgical obliteration, or both. Evaluation of feeding arteries and venous drainage as demonstrated by angiography is important when considering the appropriate treatment strategy.

**TABLE 1**

Summary of cases involving DAVFs of the anterior condylar vein and hypoglossal canal*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Presentation</th>
<th>Feeding Arteries</th>
<th>Venous Drainage</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blomquist et al., 1998</td>
<td>43, F</td>
<td>HA, pulsatile tinnitus, hypoglossal nerve palsy</td>
<td>APA, PAA</td>
<td>antegrade to epidural venous plexus</td>
<td>none†</td>
</tr>
<tr>
<td>Ernst et al., 1999</td>
<td>74, F</td>
<td>initial pulse-synchronous bruit; delayed chemosis, proptosis</td>
<td>not specified</td>
<td>initial antegrade to JV; delayed retrograde to IPS, CS, SOV</td>
<td>initial TA; delayed TV</td>
</tr>
<tr>
<td></td>
<td>74, F</td>
<td>HA, pulse-synchronous bruit</td>
<td>not specified</td>
<td>antegrade to JV; retrograde via SS &amp; TS</td>
<td>initial TA, SRS; delayed TV</td>
</tr>
<tr>
<td>Kiyosue et al., 2001</td>
<td>57, M</td>
<td>HA, pulse-synchronous bruit</td>
<td>APA, OA</td>
<td>antegrade to JB &amp; JV retrograde to IPS, CS, SOV, SPS</td>
<td>TA, TV</td>
</tr>
<tr>
<td></td>
<td>83, M</td>
<td>proptosis, chemosis, diplopia, bruit</td>
<td>APA, OA, tentorial branch of ICA, meningeal branch of VA</td>
<td></td>
<td>TV</td>
</tr>
<tr>
<td>Tanoue et al., 2005</td>
<td>70, M</td>
<td>cervical myelopathy</td>
<td>APA</td>
<td>retrograde to ASV</td>
<td>TA</td>
</tr>
<tr>
<td></td>
<td>65, F</td>
<td>pulsatile tinnitus, exophthalmos, chemosis</td>
<td>APA, OA</td>
<td>retrograde to IPS, CS, SOV</td>
<td>TV</td>
</tr>
<tr>
<td>present case</td>
<td>53, M</td>
<td>pulsatile tinnitus, hypoglossal nerve palsy</td>
<td>APA, PAA, OA</td>
<td>antegrade to vertebral venous plexus</td>
<td>TA, op</td>
</tr>
</tbody>
</table>

* APA = ascending pharyngeal artery; ASV = anterior spinal vein; CS = cavernous sinus; HA = headache; ICA = internal carotid artery; IPS = inferior petrosal sinus; JB = jugular bulb; JV = jugular vein; OA = occipital artery; PAA = posterior auricular artery; SOV = superior ophthalmic vein; SPS = sphenoparietal sinus; SS = sigmoid sinus; SRS = stereotactic radiosurgery; TA = transarterial embolization; TS = transverse sinus; TV = transvenous embolization; VA = vertebral artery.
† This DAVF was not treated because it had spontaneously thrombosed after the initial diagnostic angiogram.
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References


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