Dural Arteriovenous Fistula Associated With a Vestibular Tumor: An Unusual Case and Review of the Literature

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Abstract

Intracranial dural arteriovenous fistulae (DAVF) are rare vascular malformations. They are generally considered to be acquired lesions, often attributed to dural sinus thrombosis and intracranial venous hypertension. The authors encountered a case of DAVF associated with an octreotide-positive vestibular schwannoma. A 46-year-old female had symptoms of right ear congestion accompanied by pulsatile tinnitus and mild hearing loss. Magnetic resonance imaging (MRI) identified a lobulated mass centered at the cerebellopontine angle. Preoperatively, on cerebral angiography, there was an incidental discovery of a DAVF in the right posterior fossa. The decision was made to proceed with resection of the tumor in a staged fashion. Her latest follow-up MRI showed no evidence of recurrent tumor. This is the second reported case of DAVF associated with an intracranial schwannoma. Findings are discussed along with a thorough review of the literature. This case, combined with the data from the literature review, led us to believe that tumor-related angiogenesis might contribute to DAVF formation.

Categories: Neurosurgery
Keywords: dural arteriovenous fistula, vestibular schwannoma, octreotide scintigraphy, somatostatin receptor, angiogenesis, intracranial tumor

Introduction

Intracranial dural arteriovenous fistulae (DAVF) are abnormal direct shunts between dural arteries and dural venous sinuses, meningeal veins or cortical veins. They are located within or near the wall of the dural sinus and account for 10%-15% of all intracranial arteriovenous malformations [1]. DAVF primarily occur in adult patients and are most commonly located in the transverse-sigmoid sinus region followed by the cavernous sinus; however, any intracranial venous sinus may be involved [2]. They are widely considered to be acquired lesions. Their acquired nature is further supported by their association with dural sinus thrombosis, head trauma, intracranial or spinal tumors [3-4], localized infection, previous neurosurgical procedures [1-2], and hypercoagulation states [1].
We encountered a rare case of a transverse-sigmoid sinus DAVF associated with the presence of a vestibular schwannoma, which occluded the right transverse and sigmoid sinuses. The pre-operative diagnosis of this lesion was confounded by the fact that the tumor was positive on octreotide nuclear scintigraphy.

Case Presentation

A 46-year-old female had symptoms of right ear congestion accompanied by pulsatile tinnitus and mild hearing loss. Neurological examination revealed weakness of cranial nerves X and XII. Magnetic resonance imaging (MRI) identified a 4.2 x 4.7 x 4.1 cm lobulated mass centered at the cerebellopontine angle that was hypointense on T1-weighted, heterogeneous on T2-weighted, and avidly enhancing on post-contrast images (Figure 1A). No significant component was noted within the internal auditory canal. An Octreoscan was performed, which showed intense increased tracer uptake in the mass centered in the region of the right jugular foramen, thereby supporting the suspected diagnosis of paraganglioma (Figure 1B).

FIGURE 1: Magnetic resonance imaging and Octreoscan

T1-weighted post-contrast MRI (magnetic resonance imaging) (A) demonstrates the avidly enhancing large right skull base mass centered at the jugular foramen, involving the right
carotid space inferiorly and the right hypoglossal canal. 24-hour In-111 Pentetreotide SPECT-CT (single-photon emission computed tomography) fused images (B) demonstrates intense uptake in the mass.

Initial post-operative T1-weighted post-contrast MRI (C) demonstrates expected post-surgical changes related to the skull base mass resection with no residual enhancing tumor. Follow-up MRI (D) four years later demonstrates stable post-operative changes with no residual mass. Tiny focus of enhancement beneath the tentorium likely reflected residual DAVF (dural arteriovenous fistula) (red arrow).

Digital subtraction angiography (DSA) demonstrated a surprising lack of vascularity associated with the tumor. However, there was an incidental discovery of a Cognard Iia+b dural arteriovenous fistula in the right posterior fossa associated with an occluded right sigmoid sinus (Figure 2A-2D).

FIGURE 2: Digital subtraction angiography

Initial cerebral angiogram planned for pre-operative embolization of the suspected paraganglioma did not demonstrate significant tumor vascularity. Incidentally, a dural arteriovenous fistula was seen predominantly supplied by the right AICA (anterior inferior cerebellar artery) on right vertebral artery injection (A). The right AICA is irregular with mild fusiform dilatation (yellow arrow). Microcatheter injection in the right SCA (superior cerebellar artery) (B) demonstrates a smaller component of the fistula not well seen on the right vertebral artery injection due to rapid shunting into the AICA. Additional very small arterial feeders were identified from branches of the right MMA (middle meningeal artery). There was cortical venous drainage (blue arrow) along the right cerebellar hemisphere to the right transverse sinus (red arrow), which then drained in retrograde fashion across the torcula and into the left transverse and sigmoid sinuses.

Following embolization using Onyx in the right AICA and SCA DAVF (dural arteriovenous fistula) feeders, left vertebral artery DSA (digital subtraction angiogram) injection (C) demonstrates significant reduction in arteriovenous shunting. Again, there was no appreciable vascularity in the skull base mass. Right CCA (common carotid artery) injection (D) following embolization did not reveal additional feeders to the DAVF or mass.

Follow-up DSA four years after initial embolization. Right CCA injection (E) reveals recurrence of the DAVF with new prominent feeders from the right MMA (blue arrow) and occipital (red arrow) arteries. Following embolization using Onyx in the right MMA feeder, right CCA injection in the late arterial phase (F) demonstrates resolution in shunting through the right CCA.
Right vertebral artery injection on the most recent DSA following MMA embolization demonstrates mild residual shunting similar to prior. There has been interval resolution of the dysplastic proximal right AICA.

Due to the extensive volume of tumor, the patient underwent a staged surgical resection. The first surgery included a modified radical right neck dissection, right superficial parotidectomy with facial nerve dissection, as well as the initial approach to the tumor and partial extradural resection. One week later, the patient underwent the second stage surgery where the tumor was radically resected using a right transcoclear approach. The right sigmoid sinus was ligated. The facial nerve was mobilized and transposed. Postoperative MRI confirmed complete resection of the tumor (Figure 1C). Histological analysis of samples from both surgeries confirmed the tumor not to be a paraganglioma, but instead a schwannoma (Figure 3).

FIGURE 3: Photomicrograph

Hematoxylin and eosin stained sections showed a biphasic tumor composed of spindle-shaped cells arranged in compact interfacing fascicles with areas of denser cellularity alternating with more loosely arranged cells, consistent with a schwannoma. Tumor cells were immune-positive for S100 (not shown). Areas of collagen deposition, ancient-type changes and lymphocytic perivascular infiltrates were also present (200x magnification).

She had a long recovery from the surgery due to the lower cranial nerve palsies but has shown gradual improvement. Her latest follow-up MRI (48 months from surgery) (Figure 1D) showed no evidence of recurrent tumor; however, there was some residual posterior fossa DAVF. At that time, she also reported bothersome pulsatile tinnitus that she perceives on the right despite a complete hearing loss on the right side. DSA revealed new large feeders from the right middle meningeal artery (MMA) and occipital arteries into the residual DAVF (figure 2E), which were not present on initial presentation (Figure 2D). The DAVF was embolized utilizing Onyx in the
right MMA. Final right common carotid artery (CCA) angiogram demonstrated resolution in arteriovenous shunting (Figure 2F). Vertebral artery injection after embolization demonstrated minimal residual shunting via the right anterior inferior cerebellar artery (AICA) (Figure 2G). A further intervention was declined by the patient and angiographic follow-up in one year has been scheduled.

**Discussion**

In our patient, we encountered a transverse-sigmoid sinus DAVF associated with a schwannoma of the right skull base. Although angiography was not available prior to diagnosis of the large skull base mass, one could hypothesize that the schwannoma led to the venous sinus obstruction, and subsequently the formation of the DAVF. DAVF formation attributed to a tumor has been described, but it is rare. A detailed PubMed query was performed, which identified 26 previously reported cases (Table 1) [3-17]. Analysis of the previously reported cases exhibited a slight female predominance (61.5%) and a mean age of 60.26 years (range 25-78). This is only the second case associated with an intracranial schwannoma. In the vast majority of cases (78%), the tumor was a meningioma. Although there are exceptions, most cases of DAVF associated with tumor are also associated with sinus obstruction, underlining the importance of venous outflow obstruction in DAVF pathophysiology. Specifically, the most common location of occlusion was the sigmoid sinus, which was occluded in 38.5% of cases. Interestingly, 23% of cases were not related to sinus occlusion. The most common location for a DAVF associated with a tumor was the transverse-sigmoid sinus (48% of cases). Some investigators hypothesize that meningiomas are the most likely tumor type to be associated with DAVF due to their highly vascular nature, which in turn accentuates venous hypertension, especially when the sinus is occluded [14].

| Author          | Year  | Age/Sex | Tumor/Location                     | DAVF          | Sinus Occlusion | Treatment of DAVF | Follow-up              |
|-----------------|-------|---------|------------------------------------|---------------|----------------|-------------------|-----------------------|
| Sawamura et al. | 1991  | 68/M    | Meningioma/R occipital (parafalcine) | Tentorium     | Unclear        | Unclear (tumor surgical resection) | None reported         |
| Yokota et al.   | 1993  | 65/F    | Meningioma/R sigmoid sinus groove  | R transverse sinus | R sigmoid sinus | Pre-op TAE and surgical packing     | 36 mo (disappeared)   |
| Yamakami et al. | 1998  | 69/M    | Schwannoma/R jugular foramen       | R sigmoid sinus | R sigmoid sinus | Pre-op TAE and surgical resection   | None (disappeared post-op) |
| Arnautovic et al. | 1998 | 63/F    | Meningioma/L tentorium             | L transverse and sigmoid sinuses | L transverse sinus | Pre-op TAE and surgical resection | 40 mo (disappeared)    |
| Chung et al.    | 1999  | 57/F    | Paraganglioma/R jugular foramen    | R transverse and sigmoid sinuses | R sigmoid sinus | Pre-op TAE and surgical resection   | 28 mo (disappeared)    |
|                 |       | 62/F    | Meningioma/R tentorium             | R transverse sinus | None (tumor subtotal surgical resection) | None (disappeared post-op) |
| Authors                  | Patient ID | Diagnosis          | Vascular Anomalies | Treatment                                                                 | Follow-up |
|--------------------------|------------|--------------------|--------------------|---------------------------------------------------------------------------|-----------|
| Vilela et al. (2001)     | 51/M       | Meningioma/Parasagittal | 2 DAVFs: R superior petrosal sinus AND R parietal convexity | Surgical disconnection of DAVFs w/o tumor resection | 60 mo (no DAVF recurrence) |
|                          | 56/Unspecified | Meningioma/R skull base | R transverse sinus | None (Pre-op tumor TAE and tumor surgical resection) | 24 mo (clinically stable) |
| Horinaka et al. (2003)   | 69/F       | Meningioma/R transverse sinus | R transverse sinus and part of sigmoid sinus | Transverse sinus resection (part of tumor resection surgery) | 9 mo (disappeared) |
| Ahn et al. (2003)        | 45/F       | Meningioma/L parietal convexity | L transverse and sigmoid sinuses | None (Pre-op tumor TAE and tumor surgical resection) | 12 mo (disappeared) |
| Inoue et al. (2007)      | 76/M       | Meningioma/L sphenoidal ridge | R cavernous sinus | TVE (followed by tumor surgical resection) | 12 mo (disappeared) |
| Zhou et al. (2007)       | 42/M       | Meningioma/L anterior clinoid (tumor unnoticed pre-op) | L sphenobasilar sinus | None | Pre-op TAE and surgical disconnection | 24 mo (disappeared) |
| Toledo et al. (2010)     | 60/M       | Meningioma/L parietal lobe (parafalcine) | Superior sagittal sinus | Pre-op TAE and surgical occlusion | None (disappeared post-op) |
| Kalani et al. (2011)     | 25/F       | Hemangiopericytoma/L parieto-occipital (parafalcine) | Multiple DAVFs: Unclear locations (one torcular DAVF) | Pre-op TAE and surgical disconnection (during tumor surgical resection) | 7 mo (patient asymptomatic but has residual DAVF) |
| Enatsu et al. (2012)     | 76/F       | Meningioma/R tentorium | R transverse sinus and sigmoid sinuses | Pre-op TAE and intra-op feeder coagulation and occluded sinus resection (during tumor surgical resection) | 1 mo (disappeared) |
|                          | 55/M       | Meningioma/L sphenoid wing/orbit | 2 DAVFs: both L convexity | None (tumor subtotal surgical resection) | None reported |
|                          |            | Meningioma (radiologic) | L cavernous | | |
| Gender | Diagnosis | Sinus | Tumor Treatment | Adjuvant Treatment |
|--------|-----------|-------|----------------|-------------------|
| 73/F   | Meningioma/L occipital/tentorium | L cavernous sinus and L transverse sinus (partial) | TAE and TVE followed by GK (no tumor resection) | None reported |
| 69/M   | Meningioma/L convexity (fronto-temporal) | R transverse sinus | Partial of multiple unspecified sinuses | None (TAE of tumor followed by tumor surgical resection) | None reported |
| 41/F   | Meningioma/R lateral ventricle-trigone | L convexity | None | TAE (followed by tumor surgical resection) | None reported |
| 54/F   | Meningioma (radiologic diagnosis)/R petrous apex | L transverse and sigmoid sinuses | R cavernous sinus | TAE (followed by GK for tumor) | None reported |
| 57/M   | Meningioma (radiologic diagnosis)/L parasellar | L convexity | L cavernous sinus | None (fractionated radiotherapy for tumor) | None reported |
| 78/F   | Meningioma (radiologic diagnosis)/L convexity (frontal) | L cavernous sinus | L cavernous sinus | TAE (no tumor resection) | None reported |
| 67/F   | Phosphaturic mesenchymal tumor/L anterior cranial fossa | Lateral tentorial | None | Surgical resection (following failed TAE) | None reported |
| 72/M   | Meningioma/ Planum sphenoidale | Ethmoidal | None | Surgery (during tumor resection) | None reported |
| 61/F   | Meningioma/ Olfactory groove | R transverse and sigmoid sinuses | Superior sagittal sinus | GK and TAE | None reported |
| 70/F   | Schwannoma/ Spinal (C2 level) | R transverse and sigmoid sinuses | R sigmoid sinus | TAE and TVE (followed by tumor surgical resection). GK after DAVF recurrence 6 mo post-intervention | 33 mo (disappeared) |

**TABLE 1: Reported cases of dural arteriovenous fistulae associated with tumor in adult patients**

Another unique characteristic in our case was the positive 111In-octreotide scintigraphy (Octreoscan), which is used to evaluate the presence of somatostatin receptors in intracranial tumors in vivo. Expression of somatostatin receptors has been linked with increased angiogenic activity in meningiomas and other neoplasias [18]. This is only the second reported case of an intracranial schwannoma with positive octreotide scintigraphy [19]. Past studies have shown that somatostatin receptors are expressed in most meningiomas, paragangliomas and hemangiopericytomas, but not in schwannomas, with some investigators using octreotide...
scintigraphy for the differential diagnosis of meningioma versus schwannoma [19-20]. Our case further supports the conclusion by Dupuch et al. that no investigation is able to establish a definitive preoperative differential diagnosis between schwannoma and meningioma [19]. Taking into consideration the case we encountered, we hypothesize that aberrant angiogenesis through somatostatin receptor pathways may play a role in the pathogenesis of DAVF associated with intracranial tumors. Even though there is currently no evidence showing a direct connection between somatostatin pathways and DAVF formation, studies have shown that the presence of somatostatin receptors correlates with aberrant angiogenesis in various tumor settings [18]. Our hypothesis aims at opening new avenues of investigation that will help in better understanding DAVF pathophysiology.

Conclusions
In summary, we present an unusual case of DAVF associated with an octreotide positive vestibular schwannoma. This case demonstrates that lesions in the cerebellopontine angle could affect local vasculature, possibly by both causing local venous hypertension and somatostatin receptor-related angiogenesis, to the extent it may lead to a DAVF formation.

Additional Information
Disclosures
Human subjects: Consent was obtained by all participants in this study. Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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