CASE REPORT

Endovascular management of six simultaneous intracranial dural arteriovenous fistulas in a single patient

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SUMMARY

A 64-year-old man with a history of traumatic brain injury 4 years previously presented with progressive cognitive decline and gait abnormality. MRI revealed diffusion restriction in the bilateral centrum semiovale and multiple serpiginous flow voids. Cerebral angiogram revealed a total of six intracranial dural arteriovenous fistulas with separate fistulas of the right and left sphenoid bones, left clival plexus, right transverse sinus, right sigmoid sinus, and superior sagittal sinus. A diffuse pseudophlebitic pattern of venous drainage indicating severe venous hypertension was also observed. The patient underwent a series of endovascular treatments over the next 10 months to achieve resolution of all arteriovenous shunting. Repeat MRI showed resolution of the diffusion restriction and marked reduction in T2 vascular flow voids. The patient's clinical status improved significantly over the course of treatment, paralleling the improvement in venous hypertension.

BACKGROUND

Intracranial dural arteriovenous fistulas (dAVF) are thought to represent predominately acquired arteriovenous malformations arising where arteries pierce the dura in close proximity to venous sinuses or meningeal veins. The underlying pathophysiology has not been completely described although venous sinus thrombosis and venous hypertension play a significant role. DAVF account for 10–15% of all intracranial vascular malformations, and multiple different classification systems for these lesions have been proposed. The Borden and Cognard classification systems are the most widely used.^{1 2}

Both systems are based on the pattern of venous drainage from the fistula (table 1).

Borden types II and III along with Cognard types IIb–V represent aggressive clinical entities that warrant treatment.^{1–3} Multiple dAVF are occasionally seen simultaneously in the same patient, and different fistulas can develop over time in the same patient.^{4 5} The case presented here, to the best of our knowledge, represents the largest reported number of dAVF seen simultaneously in one patient.

CASE PRESENTATION

The patient is a 64-year-old man with a history of traumatic brain injury secondary to a high speed motor vehicle collision 4 years prior to presentation. He was hospitalized elsewhere and details of that hospitalization are not known. He presented to our institution with cognitive decline, progressive memory loss, generalized weakness, and subacute onset of gait imbalance and urinary incontinence. Neurological examination revealed confusion with orientation to person and place only. He was able to move all four extremities to commands with no focal weakness. Left-sided dysmetria and a tendency to veer left when ambulating were noted. Palmomental reflexes and Babinski signs were present bilaterally.

INVESTIGATIONS

Initial MRI showed diffuse white matter fluid attenuated inversion recovery signal abnormality, diffusion restriction in the bilateral corona radiata, and extensive enlarged serpentine vascular flow voids,

Table 1 Summary of Borden and Cognard classification systems^{1 2}

Borden classification		Cognard classification	
Type	Fistula location/venous drainage	Type	Fistula location/venous drainage
I	Venous sinus or meningeal vein(s)/antegrade in sinus(es) only	I	Venous sinus/antegrade in sinus(es)
II	Venous sinus or meningeal vein/retrograde in cortical vein(s)	IIa	Venous sinus/retrograde in sinus(es)
		IIb	Venous sinus/retrograde in cortical vein(s)
III	Cortical vein/retrograde in cortical vein(s)	IIa+b	Venous sinus/retrograde in both sinus(es) and cortical vein(s)
		III	Cortical vein (without venous ectasia)/retrograde in cortical vein(s)
		IV	Cortical vein (with venous ectasia)/retrograde in cortical vein(s)
		V	Cortical vein/retrograde in spinal perimedullary vein(s)

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Table 2 Description, classification, and treatment of each fistula

No	Fistula location	Arterial supply	Venous drainage	Borden/Cognard classification	Treatment
1	Left sphenoid wing (intraosseous)	Dural branches of OphA ILT MMA Distal I-max branches	Pterygoid venous plexus Spheno-parietal sinus	1/1	Onyx Coiling
2	Left clival plexus	Clival branch of the MHT	IPS	1/1	None, spontaneous resolution
3	Right sphenoid wing (intraosseous)	Dural branches of OphA ILT Distal I-max branches	Pterygoid venous plexus Basal vein of Rosenthal	3/3	Onyx nBCA
4	Right transverse sinus	Basal tentorial branch of MHT Transmastoid branches of OccA Muscular branches of VA	Left transverse–sigmoid sinus Straight sinus	2/2a+b	Coiling Onyx
5	Superior sagittal sinus	Anterior branch of MMA Transosseous branches of STA	Cortical subarachnoid veins	3/3	Coiling via direct puncture of SSS Onyx nBCA
6	Right sigmoid sinus	Transmastoid branches of OccA	Spinal perimedullary veins SPS	2/5	nBCA

ILT, inferolateral trunk; IPS, inferior petrosal sinus; I-max, internal maxillary artery; MHT, meningohypophyseal trunk; MMA, middle meningeal artery; nBCA, n-butyl cyanoacrylate; OccA, occipital artery; OphA, ophthalmic artery; SPS, superior petrosal sinus; SSS, superior sagittal sinus; STA, superficial temporal artery; VA, vertebral artery.

consistent with intracranial venous hypertension. Cerebral angiogram demonstrated six different dAVFs (table 2), a diffuse pseudophlebitic pattern of venous drainage indicating severe venous hypertension, partial occlusion of the mid portion of the superior sagittal sinus (SSS), and occlusion of the left internal jugular vein.

The anatomy of the six dAVF is described below. Fistula No 1 was located in the left sphenoid wing with transosseous channels from both the external carotid artery (ECA) and internal carotid artery (ICA) circulation. The arterial supply was via the dural branches of the ophthalmic artery, inferolateral trunk, middle meningeal artery, and distal internal maxillary branches. Venous drainage was mainly into the ipsilateral pterygoid venous plexus with some intracranial drainage into the ipsilateral sphenopalatine sinus (figure 1). Fistula No 2 was between a

clival branch of the meningohypophyseal trunk and the clival venous plexus with drainage into the inferior petrosal sinus (figure 2). Fistula No 3 was located in the right sphenoid wing with transosseous channels from both the ECA and ICA circulation, similar to fistula No 1. The arterial supply was via the dural branches of the ophthalmic artery, inferolateral trunk, and distal internal maxillary branches. Venous drainage was into the ipsilateral pterygoid venous plexus and also by the vein of Galen via the right basal vein of Rosenthal (figure 3). Fistula No 4 was located in the right transverse sinus. The arterial supply was from the meningohypophyseal trunk, transosseous branches of the occipital artery, and muscular branches of the vertebral artery. Venous drainage was retrograde into the contralateral transverse–sigmoid sinus and straight sinus (figure 4). Fistula No 5 was located at the SSS. The arterial supply was from the

Figure 1 Fistula No 1, left sphenoid wing intraosseous dural arteriovenous fistula (dAVF). (A, B) Anteroposterior (AP) and lateral projections of the left external carotid artery (ECA) showing the left sphenoid wing intraosseous dAVF supplied by dural branches of the ECA draining into the pterygoid venous plexus (red arrows). (C, D) Dyna-CT with contrast and Dyna-CT dual volume demonstrating the exact location of the arteriovenous shunt (blue arrows). (E, F) AP and lateral projections of the left ECA after embolization, demonstrating complete obliteration of the fistula. (F) Post-embolization Dyna-CT showing the location of the Onyx cast (red arrow).

Figure 2 Fistula No 2, left clival plexus dural arteriovenous fistula (dAVF). (A) Left internal carotid artery injection lateral projection showing a hypertrophic meningohipophyseal arterial branch supplying a clival venous plexus dAVF. (B) Follow-up angiogram 3 months later showing spontaneous resolution of the fistula.

bilateral middle meningeal artery's and transosseous branches of the superficial temporal arteries. Venous drainage was solely through the subarachnoid veins (figure 5). Fistula No 6 was located in the right sigmoid sinus. The arterial supply was from the transmastoid branches of the occipital artery and venous drainage was into a spinal perimedullary vein (figure 6). A hematology consultation was obtained, and a hypercoagulability workup was negative for any hereditary or acquired thrombophilia.

TREATMENT

Multiple different treatments were performed over the next 10 months to achieve total angiographic occlusion of all dAVF. The fistulas that posed the greatest risk to the patient (ie, highest Borden/Cognard classification) were targeted for treatment first. Thus fistula Nos 4, 5, and 6 were treated first, followed by Nos 1 and 3; fistula No 2 was not directly treated as it showed spontaneous resolution on follow-up angiograms. Percutaneous femoral arterial approaches were performed during each embolization session. Percutaneous left external jugular and direct puncture of the SSS were also utilized to embolize fistula Nos 3 and 5, respectively. Multiple different types of embolic agents, including coils, n-butyl cyanoacrylate, and Onyx, were required to abolish all arteriovenous shunting (table 2).

Figure 3 Fistula No 3, right sphenoid wing intraosseous dural arteriovenous fistula (dAVF). (A, B) Anteroposterior (AP) and lateral projections of the right external carotid artery (ECA) showing an intraosseous dAVF of the right sphenoid wing (red arrows) draining into the basal vein of Rosenthal (green arrows). (C, D) Dyna-CT and dual volume images showing the precise location of the arteriovenous fistula (blue arrows). (E, F) Post-embolization AP and lateral projections of the right ECA demonstrating complete obliteration of the fistula.

OUTCOME AND FOLLOW-UP

The patient had left-sided hemiparesis after the first embolization (partial devascularization of fistula No 4) that initially was 2–3/5 but improved to 4+/5. Otherwise the patient had no other procedure related complications and showed progressive improvement in his neurologic function over the course of treatment. At the 6 month follow-up, he was clinically asymptomatic except for mild left-sided hemiparesis. MRI performed at 6 months after the last treatment showed marked improvements. The restricted diffusion present initially in the bilateral centrum semiovale had completely resolved, and there was also a marked reduction in the serpentine vascular flow voids, indicating improvement in intracranial venous hypertension (figure 7).

DISCUSSION

Intracranial dAVF account for 10–15% of all the intracranial vascular malformations. Their etiology has not been entirely elucidated but rodent models involving creation of direct fistulas between the carotid artery and external jugular vein have shown a correlation between venous hypertension and venous sinus thrombosis with the development of new dAVF.^{6 7} Multiple intracranial dAVF have been described but are relatively uncommon. Van Dijk *et al*⁸ reported an 8.1% incidence of multiple intracranial dAVF in their large series of 284 patients with cranial and spinal dAVF. Another large series of 105 patients reported an incidence of 6.7%.⁹ Li *et al*¹⁰ reported a patient with eight intracranial vascular malformations; two of these were dAVF with the rest being aneurysms and an arteriovenous malformation. Mendonca *et al*¹¹ reported a case of rapidly progressive dementia in a patient who harbored four dAVF causing venous hypertensive encephalopathy. There are multiple other accounts of patients harboring two or more intracranial dAVF simultaneously.^{4 5 12–18} Our patient harbored a total of six dAVF on his initial angiogram. Two of these fistulas were revealed with subsequent Dyna-CT reconstructions to be intraosseous in location within both greater sphenoid wings. These intraosseous fistulas demonstrated both extracranial and intracranial venous drainage along with an arterial supply from both the ECA and ICA circulations (figures 1, 3).

This case highlights many important aspects of treating patients with multiple dAVF and intracranial venous

Figure 4 Fistula No 4, right transverse sinus dural arteriovenous fistula (dAVF). (A–C) Anteroposterior (AP) and lateral projections of the right external carotid artery (ECA) (A, B) and internal carotid artery (ICA) (C) showing a right transverse sinus dAVF supplied mainly by transmastoid branches (black thin arrow) of the occipital artery and basal tentorial artery of the ICA draining in a retrograde fashion into the transverse sinus (black arrows) and torcula. Visualization of the right sphenoid wing fistula (fistula No 3) is also demonstrated with these images (white arrows). (D–F) Post-embolization AP and lateral projections of the right ECA (D, E) and ICA (F) demonstrating complete obliteration of the fistula.

Figure 5 Fistula No 5, superior sagittal sinus (SSS) dural arteriovenous fistula (dAVF). (A, B) Anteroposterior (AP) and lateral projections of the right external carotid artery (ECA) showing the SSS dAVF (red arrows) with only cortical venous drainage (green arrow). (C) Lateral x-ray demonstrating access into the SSS by direct puncture (blue arrow). (D) Intraoperative picture of the sheath insertion directly into the SSS. (E, F) Right ECA AP and lateral projections post-embolization demonstrating complete obliteration of the SSS fistula.

Figure 6 Fistula No 6, right sigmoid sinus dural arteriovenous fistula (dAVF). (A, B) Anteroposterior (AP) and lateral injection of the right occipital artery demonstrating a dAVF in an isolated right sigmoid sinus (white arrows) draining in retrograde fashion into the superior petrosal sinus (black arrow). (C) AP injection of the occipital artery demonstrating the spinal perimedullary venous drainage (white arrow). (D, E) Complete obliteration of the fistula after embolization with Onyx 18.

Figure 7 MRI before treatment showing bilateral centrum semiovale diffusion restriction (white arrows), as evidenced by the dark apparent diffusion coefficient (ADC) map (A) and bright diffusion weighted imaging (DWI) signal characteristics (B). T2 weighted images show diffuse serpentine vascular flow voids and white matter signal hyperintensity (C). (D–F) MRI 6 months after treatment demonstrating resolution of the restricted diffusion on ADC (D) and DWI (E) sequences, with marked improvement in serpentine vascular flow voids (F).

hypertension. Firstly, it shows how cortical venous reflux and a diffuse pseudophlebitic venous drainage pattern is not only associated with a high annual risk of intracranial hemorrhage of approximately 5–10% but also with non-hemorrhagic neurologic deficits. Venous hypertension with resultant focal neurologic symptoms and/or encephalopathy can result in serious impairment.^{8 11 19–25} Our patient had experienced a severe and rapidly progressive cognitive decline secondary to chronic venous hypertension, as demonstrated on his initial MRI (figure 1). Secondly, it demonstrates how a precise understanding of each dAVF's vascular anatomy is of the utmost importance to the neurointerventionalist. The diagnosis and management of multiple intracranial dAVF is more complex than that of a single fistula in isolation. Fistulas can overlap and are difficult to visualize or distinguish from others. Multiple and selective injections are often necessary. Flat panel volume CT analysis and three-dimensional angiographic reconstructions are very helpful in understanding their complex anatomy and relation. Treatment of dAVF can result in significant improvement or even complete resolution of encephalopathy.^{19 24} Patients with severe intracranial venous hypertension and multiple dAVF require an exhaustive investigation and analysis of each individual fistula because not all of the fistulas may contribute to the venous hypertension. We suggest taking the pragmatic approach of thinking about each individual fistula in isolation, targeting the fistulas that have cortical venous reflux, and observing those that do not. Priority of treatment should go to fistulas with a higher Borden/Cognard classification. Lastly, it shows how a multidisciplinary approach is often required when managing multiple dAVF. In the current case, both endovascular and open surgical access were utilized to treat fistula No 5 (table 2). The use of multiple embolic agents, including Onyx, was also integral in treating our patient and will serve as a valuable tool in the armamentarium of any neurointerventionalist treating these complex pathologic entities.^{26 27} Cooperation and coordination of care between the neurointerventionalist and neurosurgeon is important in obtaining the best clinical outcome for these patients.²⁸

Learning points

- ▶ Multiple simultaneous dural arteriovenous fistulas are a well documented pathologic entity capable of causing severe venous hypertensive encephalopathy.
- ▶ Prioritization of treatment should go to fistula(s) with higher Borden/Cognard grades.
- ▶ Different types of embolic agents and routes of access, including direct surgical access of intracranial venous sinuses, may be necessary for adequate treatment of each fistula.
- ▶ Appropriate treatment leads to complete resolution of arteriovenous shunting and can result in dramatic radiographic and clinical improvement of the patient.

Contributors TLG (neurosurgery resident) is the primary author who contributed most substantially to the manuscript creation. LR-C (neurosurgery resident) contributed to the manuscript conception, design, and creation. CK (neurosurgery resident) contributed to the manuscript conception, design, and creation. GCR (neurologist) contributed to the manuscript conception, design, and revision. DAC (neurosurgeon) is the primary physician of record for the patient, performed open surgical access to aid in endovascular treatment, and contributed to the manuscript conception, design, and revision. OD (neurovascular interventional radiologist) performed all of the endovascular treatments for the patient and contributed to the manuscript conception, design, creation, and revision.

Competing interests OD is a consultant for Covidien (formerly ev3).

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