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Unusual Presentation of a Dural Arteriovenous Fistula of the Superior Sagittal Sinus and Single Modality Therapy with Onyx

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Superior sagittal sinus (SSS) dural arteriovenous fistulas (DAVF) are rare and present unique challenges to treatment. Complex, often bilateral, arterial supply and involvement of large volumes of eloquent cortical venous drainage may necessitate multimodality therapy. We report a case of a DAVF of the SSS in a patient who presented uniquely with increasing dizziness and disequilibrium who was treated with a single modality, endovascular embolization with ethyl vinyl alcohol co-polymer (Onyx, EV3, Irvine, CA). The patient underwent staged embolization in 2 sessions with no complications. An angiographic cure was achieved and the patient's symptoms were ameliorated. Single modality therapy with endovascular embolization of a SSS DAVF can be achieved. Careful attention to technique during embolization with Onyx is required, but complete obliteration is possible without the need for adjunctive surgical resection.

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Abbreviations: AVM, arteriovenous malformation; ACA, anterior cerebral artery; CT, computed tomography; DAVF, dural arteriovenous fistula; DMSO, dimethyl-sulfoxide; ECA, external carotid artery; ICA, internal carotid artery; MCA, middle cerebral artery; MMA, middle meningeal artery, MRI, magnetic resonance imaging; NBCA, N-butyl-cyanoacrylate

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Introduction

Superior sagittal sinus (SSS) dural arteriovenous fistulas (DAVF) are rare and present unique challenges to treatment. Complex, often bilateral, arterial supply and involvement of large volumes of eloquent cortical venous drainage may necessitate multimodality therapy.

Case Report

A 68-year-old right-handed man presented with a remote history of a right hemispheric hemorrhage of uncertain etiology at age 14 leaving him with a spastic left hemiplegia and left hemisensory loss. He had a second intracranial hemorrhage at age 21, also of unknown etiology. There was no history of trauma and no family history of hemorrhagic stroke, vascular malformation, aneurysm or bleeding diathesis. The patient was on no anticoagulants or antiplatelet therapy and there is no history of illicit drug use. In the 3 years prior to presentation, the patient was experiencing increasing dizziness, disequilibrium and loss of balance. He also complained of a longstanding history of pulsatile tinnitus and a soft pulsatile fullness in his scalp.

On examination there was an upper motor neuron pattern of left facial weakness and longstanding spastic left hemiparesis with a pyramidal pattern of weakness. Deep tendon reflexes were hyperreflexic on the left side with sustained clonus at the left ankle. There was a left hemisensory loss to primary modalities. He was able to stand and ambulate with a cane. Examination of his head and neck revealed visible pulsation of the hair on his head as well as a palpable pulsatile thrill most marked over the vertex where there was an enlarged scalp vessel.

MRI of the brain (figure 1) disclosed sequelae of previous hemorrhages with hemosiderin deposition in the right thalamus and several enlarged deep veins with a venous varix adjacent to the old thalamic hemorrhage. There were several enlarged cortical veins as well. Abnormal increased T2 signal in the pons was present. There were several enlarged vessels over the scalp bilaterally as well as in the midline at the vertex.

Catheter angiogram demonstrated a complex superior

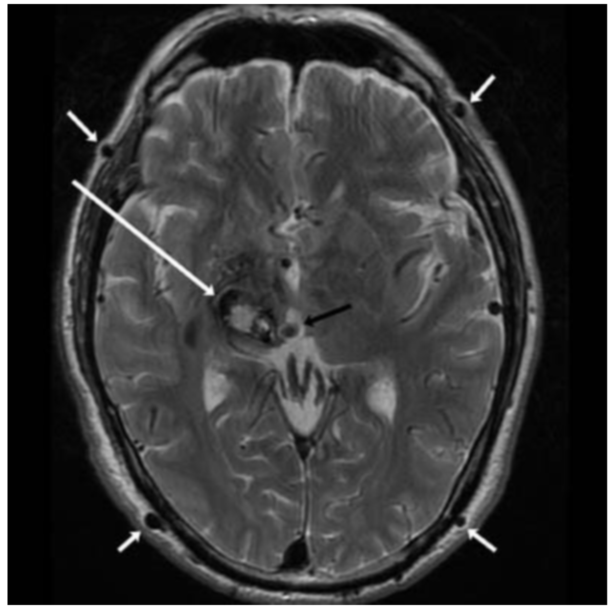

Figure 1A. 68-year-old man with a dural arteriovenous fistula of the superior sagittal sinus. Axial T2-weighted image demonstrates old right thalamic hemorrhage with hemosiderin within (large white arrow). Large vessel medially (black arrow) is a venous varix. Several flow voids are seen in enlarged scalp arteries. (Small arrows).

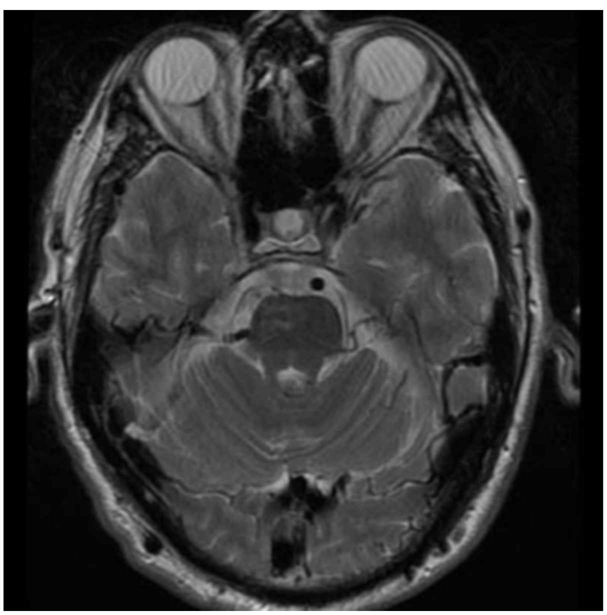

Figure 1B. Axial T2-weighted image shows abnormally increased signal in the pons.

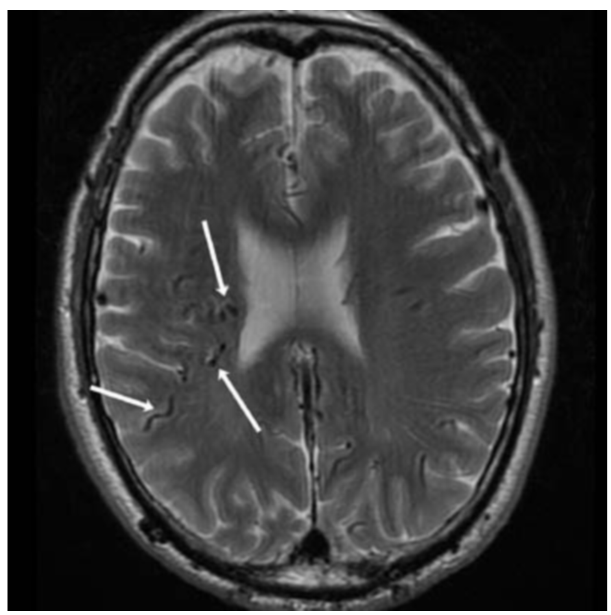

Figure 1C. Axial T2-weighted image shows several enlarged cortical veins.

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Figure 2A. Anterior-posterior image of distal right external carotid artery injection demonstrating supply to the superior sagittal sinus fistula from a large superficial temporal artery branch as well as right middle meningeal artery branches. Note filling of the superior sagittal sinus as well as several cortical veins (arrows) indicating the presence of cortical venous reflux.

Figure 2B. Lateral view of right external carotid artery injection showing both middle meningeal artery and superficial temporal artery supply to the superior sagittal sinus fistula.

sagittal sinus dural arteriovenous fistula with extensive arterial supply from branches of the external carotid arteries bilaterally, extracranial muscular branches of the left vertebral artery and pial branches of the left anterior and middle cerebral arteries (figure 2). There was cortical venous drainage seen as well.

The patient underwent endovascular therapy via a transarterial approach through the anterior division of the right middle meningeal artery (MMA). After placing a 6 French guiding catheter in the right external carotid artery, the patient was giving a loading dose of 5000 units of heparin and 1000 units per hour thereafter. A dimethylsulfoxide (DMSO) compatible microcatheter (Marathon, EV3, Irvine, CA) was manipulated over a 0.008 inch microguidewire into the anterior division of the right MMA and superselective angiography was performed. The catheter was positioned as close to the fistulous connection as was deemed safe; that is, the catheter tip was placed far enough away from the fistula so as to afford some control over preventing embolization into the superior sagittal sinus. Care was also taken to ensure that the distal tip of the catheter was in a straight segment of the MMA to facilitate catheter retrieval. The microcatheter is less likely to stick when placed in a straight rather than a tortuous segment of the vessel. The catheter was then flushed with DMSO and the dead space in the catheter was filled with Onyx 34 (8% ethyl vinyl alcohol co-polymer; EV3, Irvine, California) over approximately 40 seconds to avoid a rapid bolus of DMSO in the cerebral circulation. Higher viscosity Onyx 34 was chosen as the flow through the fistula was rapid and the higher viscosity agent was felt to less likely pass through the fistulous connection. Care was taken to avoid significant penetration into the superior sagittal sinus during the injection. Later in the course of the injection, once flow through the fistula was reduced, lower viscosity Onyx 18 (6% ethyl vinyl alcohol co-polymer) was chosen to facilitate greater penetration of the nidus of the fistula. A total of 5.7 mL of Onyx was injected over 90 minutes. The injection technique was similar to that used to treat arteriovenous malformations (AVMs) whereby a “plug” is created around the catheter tip as the Onyx forms a hardened skin with a liquid center. The plug is then pushed forward into the fistula and eventually through the fistulous con-

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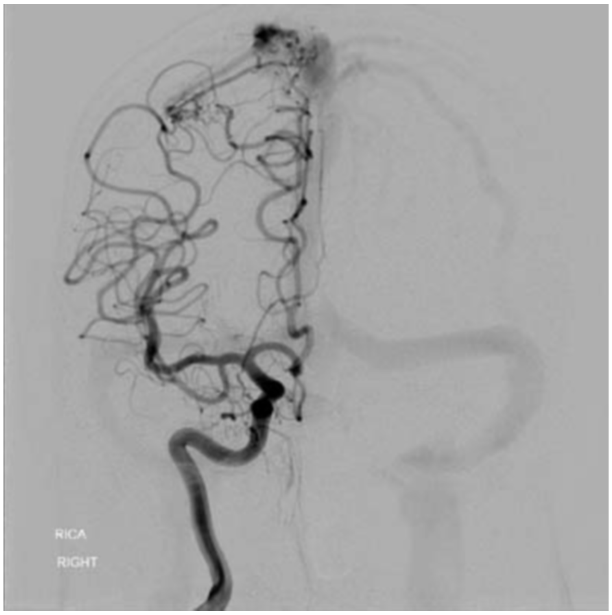

Figure 2C. AP view of right internal carotid artery injection demonstrating pial arterial supply to the fistula from branches of the right middle cerebral and anterior cerebral arteries.

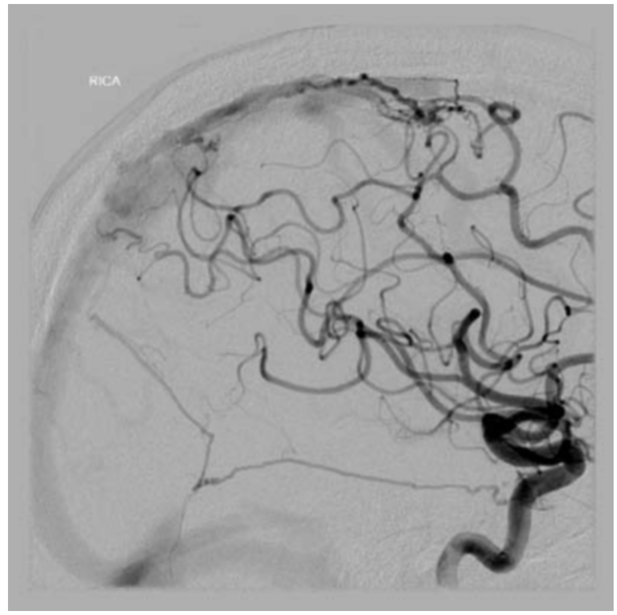

Figure 2D. Lateral view of right internal carotid artery angiogram shows meningeal branch directly from right internal carotid artery supplying fistula in addition to the middle cerebral artery and anterior cerebral artery supply.

nection. The recommended rate of injection is 0.25 ml over 90 seconds or approximately 0.16 ml per minute. The injection was stopped for 30-120 seconds when any of the three danger signs was seen: 1) penetration of Onyx into the sinus 2) when there was any reflux around the catheter tip or 3) if there was retrograde filling into another feeder, particularly if filling of a pial branch was seen (figure 2c). After waiting 30-120 seconds, the injection was continued. At this time, one often would begin to fill another portion of the fistula, often remote from the catheter tip and at a different location from the last point of opacification. Using this "waiting technique" to change the direction of filling of the fistula when a dangerous sign was identified was crucial to achieve complete obliteration of the fistula in as few microcatheter positions as possible. When restarting the injection, it was helpful to employ roadmap techniques to rapidly identify the new site of Onyx delivery to ensure a safe embolization. The endpoint was 1) reflux around the catheter tip no greater than 1.5 cm at which time the injection was terminated in order to avoid trapping of the catheter 2) inability to prevent Onyx from lodging in the sinus or 3) complete cure of the fistula.

After two sessions of embolization, the first day to treat the right side and the second day to treat the left side, the fistula was obliterated with no residual filling (Figure 3). Inadvertent retrograde filling of small right anterior cerebral artery (ACA) and middle cerebral artery (MCA) branches was not initially appreciated on fluoroscopy. This did not result in any adverse neurological outcome. The patient was seen in follow-up at six months. He experienced complete resolution of his disequilibrium, vertigo, falls, and tinnitus and no change in his longstanding spastic left hemiparesis. As well, there was no evidence of residual fistula on follow-up catheter angiogram of bilateral external carotid arteries (ECA), internal carotid artery (ICA) and vertebral arteries (Figure 3).

Discussion

Clinically this case is unique in that the patient presented with 2 remote episodes of intracranial hemorrhage at age 14 and 21. We postulate that the hemorrhages were due to venous thrombosis. The longstanding subjective bruit had been present "as long as I could remember" according to the patient indicating that the DAVF was

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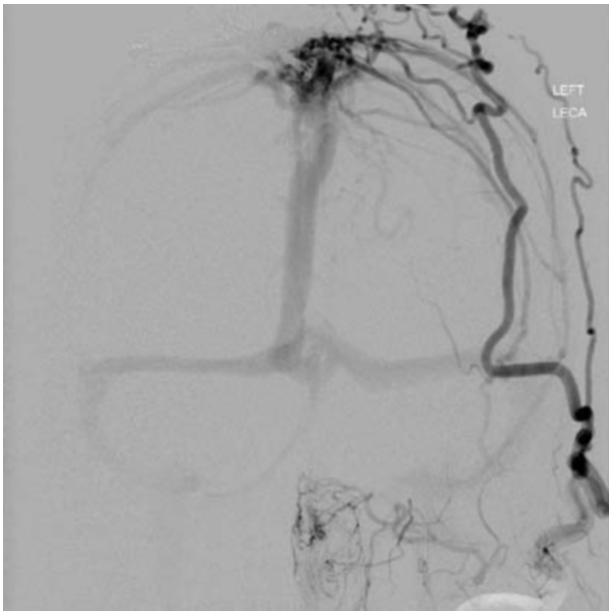

Figure 2E. After embolization of the right side, this left external carotid injection shows the left sided supply to the superior sagittal sinus fistula from the superficial temporal artery and middle meningeal artery branches.

likely present for some time, perhaps developing after his venous thrombosis. Another unusual feature of the case at hand is the repeat neurological presentation with recent symptoms of increasing dizziness, disequilibrium and loss of balance. These are unusual symptoms for a DAVF in this location. DAVF's of the SSS more commonly present with subarachnoid hemorrhage or subdural hematoma and usually during or after the fifth decade[1]. However, the signal changes in the pons may be secondary to venous hypertension related to the fistula and account for the symptoms. Similar signal changes can be seen with small vessel ischemia. However, more often there are significant supratentorial signal changes of a similar nature which were absent in this case.

Dural arteriovenous fistulas (DAVF) of the superior sagittal sinus (SSS) are rare[1]. The commonest locations are the transverse sinus, sigmoid and cavernous sinuses[1]. Superior sagittal sinus DAVF's present unique challenges to treatment due to the large volume of eloquent cortical venous drainage and the often complex bilateral arterial supply. Previously, no single modality was expected to cure a dural arteriovenous fistula (DAVF) of the superior sagittal sinus[2]. A combined approach to treatment with endovascular embolization and surgical resection is often

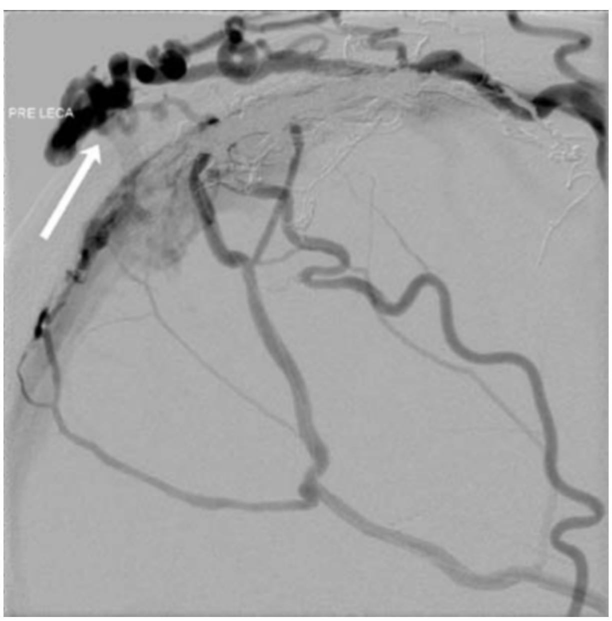

Figure 2F. Lateral view of left external carotid artery injection shows large transosseous branches of left superficial temporal artery supplying the fistula.

Figure 2G. Left vertebral artery injection shows a large muscular branch from the vertebral artery supplying the dural fistula. There was no supply from the contralateral right vertebral artery.

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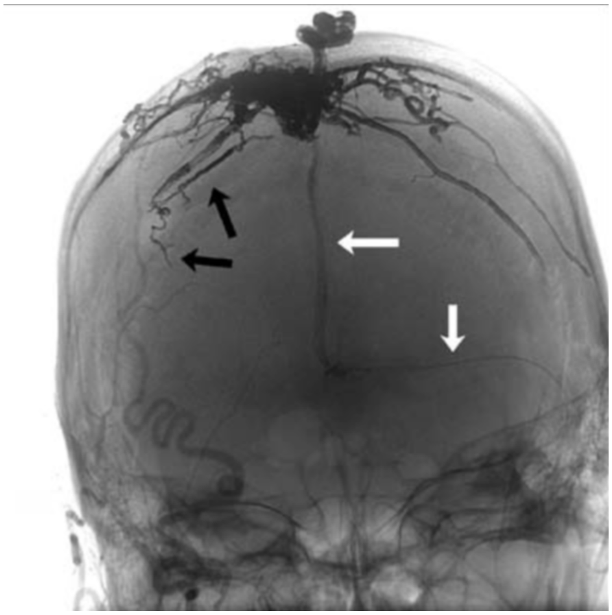

Figure 3A. Six months after treatment. Unsubtracted AP view of right external carotid artery angiogram shows Onyx cast in the fistula as well as several embolized external carotid artery branches bilaterally. Onyx cast in small cortical branches of right anterior cerebral artery (black arrows) which filled retrogradely through the fistula. Also, casting of Onyx in superior sagittal sinus and transverse sinus (white arrows) to a small degree.

required. However, resection of only the anterior 1/3 of the SSS is possible without neurological sequelae[3]. Complex DAVF's often have dural and pial arterial supply as was seen in this case, making primary endovascular cure a challenge. Furthermore, endovascular treatment with cyanoacrylates is difficult due to the fact that they are adhesive rather than cohesive and therefore limited by rapid polymerization of the glue and the propensity to occlude the superior sagittal sinus[4]. The polymerization rate, binding, and viscosity of N-butyl-cyanoacrylate (NBCA) make it an unpredictable embolic agent often resulting in proximal feeder occlusion or venous obstruction in undesirable normal territories. Onyx is a mixture of ethyl vinyl alcohol co-polymer, dimethylsulphoxide (DMSO) and tantalum[5]. Onyx is available in low concentrations of 6% and 8% for treatment of AVM's and higher 20% concentration for treatment of aneurysms. Onyx is delivered with greater control because it is non-adhesive or cohesive allowing prolonged injections and better penetra-

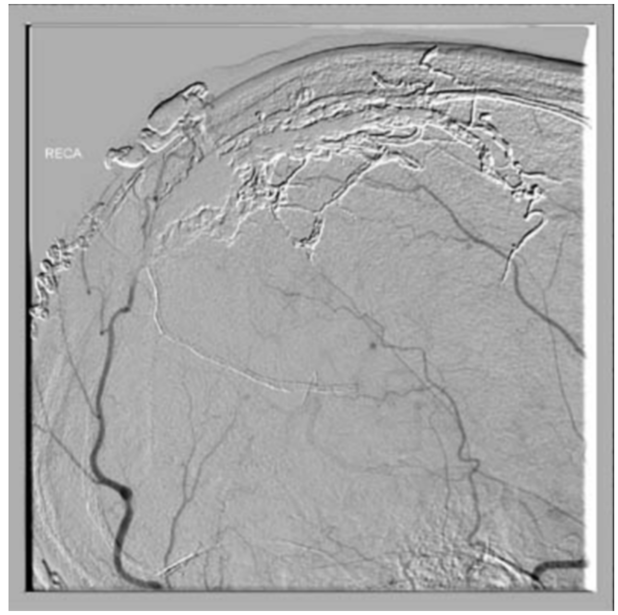

Figure 3B. Right external carotid artery lateral subtracted angiogram does not demonstrate any filling of the fistula.

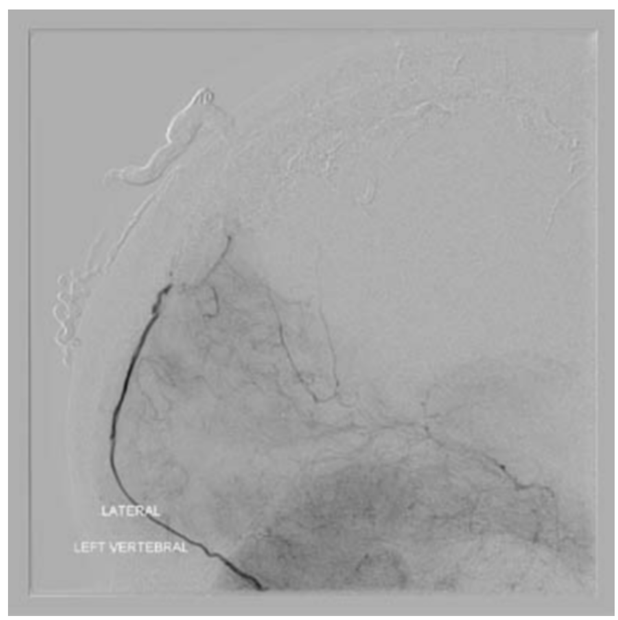

Figure 3C. Left vertebral lateral angiogram shows slow filling of the muscular vertebral artery but no filling of the fistula.

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tion[6]. Onyx does not polymerize rather it precipitates as the dimethylsulfoxide (DMSO) diffuses. The physical properties of Onyx allow good penetration of a nidus of an arteriovenous malformation (AVM)[5]. Therefore it would seem reasonable to apply these characteristics of Onyx to the treatment of complex DAVF, especially those with a nidal component. The use of Onyx in this case and the resultant cure of a large complex DAVF of the SSS are attributed to the properties of the embolic agent.

There are reports of spinal DAVF's treated with Onyx but only one other case report of DAVF of the superior sagittal sinus treated with this agent as well as reports of treatment of single cases involving the cavernous sinus and greater wing of the sphenoid [7-10] . DAVFs are fed by external carotid dural supply and rarely pial supply as seen with the present case [1]. External carotid supply has an advantage of tolerating more reflux around the microcatheter without the danger of occluding blood supply to normal brain [9]. This case and the other reported treatment of a DAVF of the SSS demonstrate that one is able to fill feeders retrogradely. Care must be taken not to fill pial feeders retrogradely thereby risking compromise of blood supply to normal brain. This requires diligent monitoring of the active site of opacification with Onyx which can be remote from the tip of the microcatheter. Reflux of Onyx retrogradely into a single anterior cerebral artery branch feeding the DAVF was seen in this case, without neurological sequellae. Notably, Arat et al demonstrated that pial feeders can regress after embolization of the DAVF with Onyx [9]. The long term effectiveness of Onyx for treatment of DAVF's requires further investigation. In a case reporting the use of Onyx to treat a carotid cavernous fistula, the authors saw no recanalization at 3 months post embolization [8]. Our case showed no recanalization at 6 months follow-up angiography.

References

1. Kurl S, Saari T, Vanninen R, Hernesniemi J. Dural arteriovenous fistulas of superior sagittal sinus: case report and review of literature. Surg Neurol. 1996 Mar;45(3):250-5. [PubMed]

2. Bertalanffy A, Dietrich W, Kitz K, Bavinzski G. Treatment of dural arteriovenous fistulae (dAVF's) at the superior sagittal sinus (SSS) using embolisation combined with micro- or radiosurgery. Minim Invasive Neurosurg. 2001 Dec;44(4):205-10. [PubMed]

3. Kobayashi H, Hayashi M, Noguchi Y, Tsuji T, Handa Y, Caner HH. Dural arteriovenous malformations in the anterior cranial fossa. Surg Neurol. 1988 Nov;30(5):396-401. [PubMed]

4. Howington JU, Kerber CW, Hopkins LN. Liquid embolic agents in the treatment of intracranial arteriovenous malformations. Neurosurg Clin N Am. 2005 Apr;16(2):355-63, ix-x. [PubMed]

5. Jahan R, Murayama Y, Gobin YP, Duckwiler GR, Vinters HV, Vinuela F. Embolization of arteriovenous malformations with Onyx: clinicopathological experience in 23 patients. Neurosurgery. 2001 May;48(5):984-95; discussion 95-7. [PubMed]

6. Murayama Y, Vinuela F, Ulhoa A, Akiba Y, Duckwiler GR, Gobin YP, et al. Nonadhesive liquid embolic agent for cerebral arteriovenous malformations: preliminary histopathological studies in swine rete mirabile. Neurosurgery. 1998 Nov;43(5):1164-75. [PubMed]

7. Warakaulle DR, Aviv RI, Niemann D, Molyneux AJ, Byrne JV, Teddy P. Embolisation of spinal dural arteriovenous fistulae with Onyx. Neuroradiology. 2003 Feb;45(2):110-2. [PubMed]

8. Arat A, Cekirge S, Saatci I, Ozgen B. Transvenous injection of Onyx for casting of the cavernous sinus for the treatment of a carotid-cavernous fistula. Neuroradiology. 2004 Dec;46(12):1012-5. [PubMed]

9. Arat A, Inci S. Treatment of a superior sagittal sinus dural arteriovenous fistula with Onyx: technical case report. Neurosurgery. 2006 Jul;59(1 Suppl 1):ONSE169-70; discussion ONSE-70. [PubMed]

10. Rezende MT, Piotin M, Mounayer C, Spelle L, Abud DG, Moret J. Dural arteriovenous fistula of the lesser sphenoid wing region treated with Onyx: technical note. Neuroradiology. 2006 Feb;48(2):130-4. [PubMed]