CASE REPORT

Combined Arterial and Venous Approaches for Cure of Carotid–Cavernous Sinus Fistula in a Patient with Fibromuscular Dysplasia

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Abstract—We report the case of a woman with bilateral carotid artery fibromuscular dysplasia. The case illustrates some of the difficulties encountered in the treatment of carotid–cavernous fistula. For example, a previous contralateral carotid closure caused complications because carotid sacrifice was not now possible. This resulted in several embolization procedures requiring prolonged x-ray exposures. We show that transvascular embolotherapy must incorporate a flexible approach for access, selection of embolic materials, and clinical care. (*Skull Base Surgery*, 4(2):103–109, 1994)

Fibromuscular dysplasia (FMD) frequently affects the internal carotid artery (ICA) and may be associated with intracranial aneurysm, thromboembolism, and arterial dissection. When dissection involves the intracavernous segment of the ICA, a fistula may develop between the artery and the cavernous sinus (carotid–cavernous fistula [CCF]).^{1–3} FMD may predispose to the development of bilateral fistulas, which present added therapeutic difficulties.^{4,5} The following case study illustrates some of the difficulties involved in treating such patients and how transvascular embolotherapy must incorporate a flexible approach for access, selection of embolic materials, and clinical care. Advances in interventional neuroradiology may permit cure in cases previously considered to be incurable with more standard surgical or interventional techniques.

CASE REPORT

A 29-year-old black woman with a long history of bilateral carotid artery FMD presented to her local emergency room with moderate bilateral proptosis (right > left), diplopia, blurred vision, and pulsatile tinnitus. There were no abnormal mental status findings. Eight years prior

to this admission, a left-sided CCF had been discovered and obliterated by a surgical trapping procedure, which required sacrifice (occlusion) of the left ICA by clipping the cervical and supraclinoid segments.

Angiography on admission revealed persistent occlusion of the previously trapped left ICA and a right-sided CCF, with marked irregularity and multifocal stenosis of the proximal cervical right ICA (RICA) (Fig. 1). Venous drainage from the fistula was directed posteriorly toward the inferior petrosal sinus, inferiorly toward the pterygoid venous plexus, and anteriorly toward a mildly distended right ophthalmic venous system (Fig. 2). There was no evidence of visual loss, and no therapy was offered, given the risks involved (prior carotid sacrifice and poor vascular access).

Three and a half weeks later, the patient's condition acutely deteriorated. She simultaneously developed severe right-sided headaches and increased right proptosis. She was again seen at a local emergency room, where a mental status examination revealed decreased cognitive function. Due to a lack of available local facilities, she was transferred to our institution for neuroradiological intervention.

On arrival, she was drowsy but arousable and complained of severe headache. Physical findings included

Figure 1. Selective injection of the right internal carotid artery, showing irregular stenosis (arrows), and a fenestration (long arrow) related either to the underlying fibromuscular dysplasia or to developmental variation.

bilateral proptosis (right > left), blurred vision, diplopia, and fine nystagmus on right lateral gaze. A loud audible bruit was present over the right lateral temporal region.

During the first angiographic procedure, performed on the morning of the patient's admission to the hospital, angiography revealed a tortuous, stenosed, and dysplastic cervical RICA. A portion of the artery had the classic "string of beads" appearance, characteristic of FMD. There were segments with luminal constriction between normal-appearing segments, and the artery also had a fenestration, representing either a chronic dissection or developmental variation, at the C2 level (Fig. 1). A single hole CCF was again delineated; however, when compared with the initial angiographic study, the venous drainage pattern had dramatically changed (Figs. 2 and 3). In the interim period coinciding with clinical worsening, the venous drainage toward the pterygoid plexus had thrombosed, and venous flow had been redirected toward sylvian cortical veins and the sphenoparietal sinus, a drainage pathway not detected on the previous angiogram. Since this cortical venous drainage now placed the patient at risk for the development of subarachnoid hemorrhage or venous infarction,⁶ and also likely accounted for the recent development of severe right-sided headaches, acute intervention was planned. Redirection of flow also resulted in further distension of the superior ophthalmic venous system, accounting for the observed worsening of the patient's ocular symptoms, a further reason for intervention.

The day after the first angiographic procedure, the patient returned for a transarterial approach to the CCF via the RICA. A superselective angiogram performed with a variable stiffness microcatheter (Tracker 18, Target Thera-

Figure 2. Drawn representation of the lateral view of the right internal carotid artery (RICA) angiogram in the reported case illustrating four stages of fistula closure. A: Initial outside right internal carotid arteriogram, performed after the development of right orbital findings. Note the fenestrated dysplastic RICA (1) and the abnormal venous drainage from the fistula, including the relatively small ophthalmic vein (2), the inferior petrosal sinus (3), and the patent pterygoid venous plexus (4) that courses inferiorly through the skull base. B: Right internal carotid angiography performed shortly before embolization, after the onset of severe headache and worsening of ocular signs of venous hypertension. Note interval thrombosis of the pterygoid plexus (not shown), enlargement of the ophthalmic vein (2), and newly developed drainage toward cortical middle cerebral veins via the sphenoparietal sinus (5). C: Same view after the transvenous deposition of platinum fiber coils (corkscrew) in the superior compartment of the cavernous sinus resulting in occlusion of cortical venous drainage, protecting the brain. Headaches resolved, but orbital signs worsened (represented by further enlargement of the ophthalmic vein (2)). D: Complete fistula closure following transarterial coil and tissue adhesive deposition within the inferior compartment of the cavernous sinus.

peutics, Fremont, CA), revealed that the cavernous sinus was partitioned into inferior and superior compartments (Fig. 4). Arterialized blood reaching the superior compartment drained toward the inferior petrosal sinus and the superficial sylvian cortical venous system. The inferior compartment drained exclusively into the ophthalmic venous system. Shortly thereafter, the catheter position was lost and further attempts to regain it were unsuccessful. Although CCFs are often effectively treated by sinus occlusion by transarterial delivery of a detachable balloon device,⁷⁻⁹ the high risk of inadvertent balloon detachment

Figure 3. A: Lateral view of right internal carotid artery angiogram performed after initial outside presentation, corresponding to Figure 2A (ophthalmic vein [2], inferior petrosal sinus [3], pterygoid plexus [4]). B: Lateral view shortly before embolization, demonstrating interval thrombosis of the pterygoid venous plexus, enlargement of the ophthalmic vein (2), and the newly opened cortical venous drainage (5), corresponds to Figure 2B.

Figure 4. Selective variable stiffness catheter angiography of the carotid-cavernous fistula (CCF), performed prior to treatment. Although this catheter position was obtained for the study, it could not be reobtained until the final phases of the embolization procedure. A: Catheter advanced past the fistula into the native distal cavernous segment of the internal carotid artery beyond it. B: Slight withdrawal of the catheter from the position illustrated in A, resulted in passage of the catheter tip into the cavernous sinus, through the fistula hole. Contrast injection resulted in a selective cavernous sinus venogram, filling all of the routes of venous drainage from the fistula, including the ophthalmic vein (2), inferior petrosal sinus (3), and the sphenoparietal sinus-cortical veins (5), (compare with Figures 2B and 3B). Note compartmentalization of the superior (arrow) and inferior (open arrow) divisions of the cavernous sinus.

related to the observed proximal carotid artery stenoses prevented its use for fistula closure in this case.

Because of the acute need to occlude the cortical venous drainage, which was the source of intracranial venous hypertension, the internal jugular vein was accessed via the femoral vein, and a variable stiffness catheter was placed into the jugular bulb and then into the right inferior petrosal sinus. The superior compartment of cavernous sinus was accessed by gentle manipulation of the guide wire through a small region of resistance at the junction of the petrosal sinus and the cavernous sinus. Selective cavernous sinus venography revealed filling of the sphenoparietal sinus and cortical veins, without filling of the ophthalmic venous system. The small catheter was then advanced anteriorly within the cavernous sinus to a point near its junction with the cortical veins (Fig. 5), and four 4 mm platinum fiber coils (Target Therapeutics) were positioned at the sphenoparietalcortical venous junction. Although immediate postcoil venography revealed continued slow filling of the cortical veins, the patient noted almost immediate decrease in the intensity of her right-sided headache. The final angiogram revealed occlusion of the anterosuperior cavernous sinus and obliteration of abnormal flow toward the cortical veins (Figs. 2C and 6).

Contrast injection through an arterial catheter demonstrated persistent filling of the fistula toward the remaining inferior cavernous sinus compartment (Fig. 2C). The ophthalmic vein had increased in caliber, due to redirection of flow caused by interval thrombosis of the anterosuperior cavernous sinus. Despite further efforts to access

Figure 5. Transvenous catheterization of the sphenoparietal sinus-cortical venous drainage of the fistula (lateral view of venogram, anterior toward the left). Note the base catheter in the jugular bulb (A) and the course of the variable stiffness catheter (small arrows) through the superior compartment of the cavernous sinus, with its tip clearly marked (arrow).

the residual fistula by arterial catheterization, spasm developed, preventing further attempts at embolization at that time.

During the 48-hour period following the procedure, the patient reported a marked decrease in the intensity of her right-sided headache; however, blurred vision and diplopia persisted. Nystagmus was present in all fields, and there was increased scleral redness. Proptosis had slightly diminished, and the patient was now able to close her eyes fully. However, on the third day following the first intervention, the patient's condition deteriorated. She reported severe right orbital pain and on examination there was increased proptosis. In light of the patient's deteriorating condition, and despite prior failure, it was decided to embolize the remaining fistula urgently.

The following morning, another interventional procedure was performed. A Tracker 18 catheter was, this time, successfully navigated through the fistula into the cavernous sinus. There appeared to be no significant change in the venous drainage pattern from that present after the successful exclusion of the cortical venous drainage. At this point, several unsuccessful attempts were made to occlude the fistula and the remaining venous drainage. An initial attempt to embolize the fistula with platinum fiber coils resulted in the successful placement of only one coil. It was elected to terminate the interventional procedure, and the patient's right orbital pain was treated with analgesics.

Over the following 3 days, the patient's eye symptoms became worse. The right eye remained severely proptotic and painful. The patient also experienced severe right orbital pain. Morphine administration provided moderate pain relief. Extraocular motion was impaired, with marked restriction of right lateral gaze. The patient complained of continued diplopia and blurred vision.

The next day another attempt was made to embolize the CCF. Angiography revealed a venous drainage pattern within the cavernous sinus that was no different from that seen 3 days earlier (Fig. 6B). At this point, a variable stiffness catheter was advanced through the RICA, although it could not be placed more than 1 cm through the CCF into the cavernous sinus. The prior introduction of coils had partly stenosed its lumen, preventing the catheter from being advanced any further. Slow injection of contrast material into the sinus revealed filling of its lumen, without any reflux of contrast into the RICA. The lack of contrast reflux into the arterial system prompted an attempt to embolize the remainder of the fistula with the injection of 0.1 cc of Avacryl. The patient experienced a moderate amount of local pain during the injection procedure; however, angiography revealed successful closure of the CCF and complete obliteration of abnormal venous flow, with preservation of normal antegrade flow within the intracranial territory of the ICA (Fig. 7). Marked irregularity of the precavernous segment of the RICA was evidence of the arterial dissection that was the likely etiology of the fistula. Over the next several days, all

Figure 6. Scout image showing coils (A) and lateral view, right carotid angiogram (B) after obliteration of cortical venous drainage by coil placement (ophthalmic vein [2], internal carotid artery [1], inferior petrosal sinus [3]), corresponds to Figure 2C.

orbital symptoms resolved. The patient remains neurologically intact to this date.

DISCUSSION

This case illustrates several important features of CCF. From a clinical perspective, the relationship between

the changing pattern of venous drainage and signs and symptoms is well demonstrated. On initial presentation, angiography revealed venous drainage toward the ophthalmic vein, petrosal sinus, and the pterygoid venous plexus. The ophthalmic vein was relatively small, correlating with the observed low level of proptosis and conjunctival changes. The delayed onset of severe headache and the acutely increased proptosis corresponded to the redirection of venous drainage toward the cortical and ophthalmic veins, respectively, following spontaneous thrombosis of the pterygoid venous drainage pathway (demonstrated by the change in venous drainage pattern between angiograms, Figs. 2 and 3). The severe headache was clearly related to cortical venous hypertension, and it persisted until selective closure of the sylvian venous drainage with coils during the first stage of embolization (Fig. 6C). Selective closure of the cortical veins by a venous approach was performed first to protect the brain from venous hypertension and possible hemorrhage or venous thrombosis. This resulted in a predictable worsening of orbital signs and symptoms, due to a reduction in alternate cavernous sinus outlets. This risk was accepted because proximal dissection of the dysplastic internal carotid artery prevented transarterial access to the fistula site with a detachable balloon, and an initial attempt to close the fistula completely via the carotid artery had failed.

The severity of the orbital findings following cerebral protection by the transvenous closure of the superior compartment of the fistula prompted a final, prolonged, but successful attempt to close the fistula transarterially through a variable stiffness catheter. A single releasable coil could be successfully placed into the cavernous sinus; however, this was insufficient to close the fistula. Closure

Figure 7. Final selective right internal carotid angiogram following embolization (corresponds to Figure 2D). Note residual irregularity of the precavernous segment of the internal carotid artery (arrow), related to underlying fibromuscular dysplasia.

was finally accomplished by the injection of a small amount of acrylic glue, a rapidly polymerizing embolic material that has been used for occlusion of fistulas in selected cases.^{10,11} A relatively high-risk glue injection technique was required (with positioning of the catheter tip near the fistula hole and the internal carotid lumen) because of a lack of good alternatives. Carotid sacrifice to close the fistula was not a viable alternative because of a previous contralateral carotid closure. The use of individually releasable coils or glue might have been unnecessary if a controlled release coil, such as the investigational Guglielmi detachable coil (GDC), had been available at the time.

This case also illustrates the risk of treating CCF complicating FMD by nonselective carotid artery occlusion and fistula trapping. Although successful for treating the contralateral fistula 8 years previously, the arterial sacrifice complicated the treatment of the second fistula. Carotid sacrifice should not be considered unless all less destructive interventional options have been fully explored. Arterial sacrifice should rarely be necessary in this era of ever-improving transvascular therapeutic alternatives.

The need to preserve the ipsilateral carotid artery in this case required extensive efforts and several embolization procedures, resulting in, among other things, prolonged x-ray exposures from digital subtraction angiography and fluoroscopic guidance of catheter positioning and embolization.¹² Transient hair loss was noted several weeks following these procedures, likely related to a depilatory dose of x-ray given to a localized area of scalp that served as the entry site for the prolonged monitoring needed to guide so many attempts at navigating the stenotic carotid artery. In the future, the length of the proposed procedure and the possible delayed effects of a radiation dose that could extend into a therapeutic dose range must be considered when judging the relative benefits and risks of x-ray-guided intervention versus other possible (usually surgical) alternatives.

We have demonstrated the utility of a flexible approach to intravascular treatment of a difficult skull base fistula. New techniques employing variable stiffness microcatheters are valuable additions to the other methods previously employed, including direct surgical approaches to the cavernous sinus.^{5,13,14} Although there are certainly potential risks involved with these procedures,¹⁵⁻¹⁷ when performed expertly the transvascular approach offers an elegant solution with potential advantages that include reduced hospital stay as a result of rapid postoperative recovery (no craniotomy).

Finally, the clinical effects of various patterns of venous drainage related to CCF can be determined by close attention to angiographic changes, as they were in this case, where an episode of spontaneous venous thrombosis resulted in a clear-cut constellation of interrelated clinical and angiographic findings.

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REVIEWERS' COMMENTS

This article makes two important points. The difficulty of this case helps illustrate the innovation that has come to interventional radiology as newer catheters and more aggressive approaches to various pathologic conditions develop. The necessity of producing highly skilled interventional neuroradiologists capable of performing these intricate tasks is obvious. A second point deserves comment. As capabilities expand, the interventional radiologists and skull base surgeons must work together. The team must recognize the capabilities of intravascular approaches, but also must protect potential access vessels during surgical intervention. The surgeons should be aware that unrelated pathologic states developing later in life may require these potential ports of approach. The authors are to be congratulated for their innovative therapy as well as their clear description. The readers should take away not only the appreciation of this interesting case, but also an appreciation of the rapidly advancing capability of interventional neuroradiology.

Hugh Curtin, M.D.

The authors should be congratulated for their persistence and skill in treating a difficult carotid–cavernous fistula (CCF). The transvenous approach permits an excellent access to recalcitrant fistulas that fail the more conventional transarterial approaches. We have utilized this approach for more than 100 cases of CCFs (both direct and indirect) with excellent results. Our experience has taught us the importance of delivering the platinum embolic device initially into the draining compartments that are least accessible from that route. For example, from a postero-inferior petrosal sinus access, it is most desirable to occlude the distal (anterior) superior ophthalmic vein and cortical venous drainage first and then occlude the fistula site primarily with coils. This was not technically possible in this case, but the authors wisely chose to close the cortical venous drainage from this access. Cortical venous drainage carries a high risk of hemorrhage and neurological deficits and should demand urgent therapy. It is interesting to note that the initial and second arteriograms (Figure 3A and B) disclosed cortical venous drainage from the fistula into the posterior fossa veins. This probably accounts for the patient's nystagmus, an unusual finding in patients with CCFs. The fistula was finally closed with a transarterial injection of a rapidly polymerizing liquid adhesive, which the authors acknowledge as a high-risk procedure. Their extensive experience with this agent was undoubtedly responsible for the excellent outcome. We and others¹ have utilized the controlled-released coil (GDC) in recent similar cases as an alternative way to treat such fistulas from both the arterial and venous routes. This should prove to be a safer and more effective technique for such difficult fistulas.

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