CASE REPORT

Segmental upper mid-basilar artery sacrifice in a child using a Micro Vascular Plug device for treatment of a basilar arteriovenous fistula compressing the brainstem

Alfred P See,¹ Michael A Kochis,² Darren B Orbach²

¹Department of Neurosurgery, Brigham and Women's Hospital, Boston, Massachusetts, USA
²Neurointerventional Radiology, Boston Children's Hospital, Boston, Massachusetts, USA

Correspondence to

Dr Darren B Orbach, Neurointerventional Radiology, Boston Children's Hospital, 300 Longwood Avenue, Boston, MA 02115, USA; darren.orbach@childrens.harvard.edu

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SUMMARY

This case demonstrates a rare pathology of basilar artery pial arteriovenous shunting presenting with acutely progressive symptoms of brainstem compression, in which we applied a polytetrafluoroethylene-coated embolic device to achieve short-segment upper mid-basilar artery sacrifice. In addition to presenting a rare form of basilar arteriovenous fistula (AVF) (likely related to hereditary hemorrhagic telangiectasia) and discussing the extreme management challenges, this case demonstrates the unusual but successful application of a novel neurovascular device. Unanswered questions include the mechanism of development of this pathology and the optimum post-embolization anticoagulation plan.

BACKGROUND

Reports of pial arteriovenous fistulas (AVFs) that occur in the posterior fossa are quite rare, occasionally appearing in general pial AVF case series or as individual case reports;^{1–6} in particular, direct AVFs originating off the basilar artery have not previously been reported. We present the case of a child with a basilar pial AVF with mass effect and progressively accelerating neurological decline and report on its management and outcome.

CASE PRESENTATION

A 3-year-old child with a history of ocular albinism and normal neurological milestones (walking at

10 months, speaking in full sentences) was noted to have several months of progressive gait instability, bilateral hand tremor, dysarthria, drooling, and intermittent esotropia, with accelerating worsening over the most recent 3 weeks. Her parents and three siblings were all healthy.

On examination she had left eye intorsion and limitation of upgaze. She ambulated with a hyperplantarflexed gait and became progressively unsteady on her feet, with axial ataxia. She had intention tremors bilaterally in the upper extremities. Over the course of observation of several days she developed swallowing difficulties and, as a result, had her oral intake progressively restricted, and showed progressive loss of motor and speech milestones.

INVESTIGATIONS

On MRI brain she was found to have an AVF that appeared to originate directly off the upper mid-basilar artery, with a massive prepontine varix compressing the right pons as well as the cerebral aqueduct, resulting in ventriculomegaly (figure 1A–C).

She was then evaluated with digital subtraction angiography 1 week later, which demonstrated a posterior wall basilar artery supply directly into the dilated venous varix (figure 2); there was no appreciable branch contribution from the posterior cerebral arteries or the superior cerebellar arteries. The

Figure 1 (A) Axial T2-weighted MRI demonstrating a pontomesencephalic high-flow arteriovenous fistula with a dilated venous varix (asterisk) harboring an arterial-signal flow void and compressing the pons and midbrain posteriorly, from left to right. (B) Sagittal contrast-enhanced T1-weighted MRI demonstrating the basilar artery (arrow) with posteriorly projecting direct flow into the pontomesencephalic venous varix. (C) Time-of-flight three-dimensional MR angiogram image in lateral projection. This demonstrates the relationship between the venous varix and the posterior cerebral artery candelabra.

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Figure 2 (A) Frontal and (B) lateral projections of a right vertebral artery injection demonstrating the dilated venous varix (asterisk) from the basilar arterial fistula. (C, D) Lateral projections of the right and left internal carotid artery injections demonstrating the robust posterior communicating artery (arrow) bilaterally and lack of arterial supply to the arteriovenous fistula (AVF) from branches other than the basilar artery. (E, F) Frontal projections of microcatheter navigation into the proximal right P1 and at the basilar apex, demonstrating the absence of AVF opacification beyond the upper mid-basilar artery. A robust network of basilar bifurcation perforators is seen at the apex (arrow).

posterior communicating arteries opacified robustly bilaterally, with arterial outflow directed toward the AVF during internal carotid artery injections bilaterally (figure 2C, D).

TREATMENT

Following multidisciplinary discussion, the decision was made to occlude the segment of basilar artery supplying the AVF. The patient was brought back 5 days later for planned embolization using a Micro Vascular Plug (MVP) device, with 81 mg oral aspirin initiated 3 days prior. Bilateral femoral access with 5 F Glidesheath Slender sheaths (Terumo Interventional, Somerset, New Jersey, USA) was achieved for catheterization of the left internal carotid artery and left vertebral artery, each with a 5 F Sofia catheter (Microvention, Tustin, California, USA). A 5 mm MVP (Covidien Medtronic, Minneapolis, Minnesota, USA) was deployed within the upper mid-basilar artery (figure 3A, B). With the device in place but before detachment, the two Sofia

catheters were used to perform angiography via the left internal carotid artery, right internal carotid artery and the lower basilar artery (figure 3A–D). The device immediately decreased arteriovenous shunting, with trace filling of the venous varix, while preserving robust supplies from the left internal carotid artery to the left posterior cerebral artery candelabra and from the right internal carotid artery into the right posterior cerebral artery candelabra and basilar apex. Smaller foci of slow arteriovenous shunting, spatially non-contiguous with the varix, continued to be seen via perforators off the left lateral basilar artery and the left anterior inferior cerebellar artery.

OUTCOME AND FOLLOW-UP

MRI on the evening following the procedure demonstrated no appreciable residual arteriovenous flow into the varix and marked improvement in pontine compression (figure 4A). There was no evidence of diffusion restriction to suggest post-embolization ischemia. She continued to have axial and appendicular ataxia but was newly awake, alert and interactive. She was continued on heparin anticoagulation with a partial thromboplastin time (PTT) goal of 60–80 for 2 days postoperatively, in addition to 81 mg aspirin daily. She was then transitioned to low molecular weight heparin and continued aspirin. Over the course of the next several days there was steady improvement in her swallowing, eye movements and ataxia. Follow-up MRI at 2 weeks showed a significant further decrease in mass effect from the thrombosing venous varix (figure 4C, D), as well as a focal right thalamic T2 hyperintensity without diffusion restriction.

One week postoperatively the patient presented with headaches, but these improved following discontinuation of the low molecular weight heparin and initiation of a 6-day prednisone taper. At 3-week clinic follow-up the parents reported that she could run and jump with no inhibition and eat without evidence of aspiration. At 3 months her parents reported her to have returned to baseline range of activity and function, although she did have persistent migrainous headaches. On formal neurological examination she had mild residual tremor and hyper-reflexia. Three-month angiographic follow-up demonstrated robust posterior cerebral supply from the internal carotid arteries, with no residual opacification of the embolized fistula (figure 4E, F). There was persistence of small foci of slow-flow arteriovenous shunting supplied by the left anterior inferior cerebellar artery and the left-sided basilar perforator, somewhat diminished compared with prior studies. On genetic investigation the patient was found to have duplication of an entire exon in the endoglin gene, implicated in hereditary hemorrhagic telangiectasia.

DISCUSSION

This case presents an unusual pathology—AVF with direct supply from the basilar artery—discovered in the setting of acute clinical decline attributable to mass effect from the dilated draining venous varix.

Open surgical approaches would be complicated by the direct fistulous connection between the basilar artery and the massive varix. The direct single-hole fistula from the basilar artery to the venous pouch and the absence of any intervening feeding pedicle between the basilar artery and the varix made targeted coil embolization (as was performed by Yoshida *et al.*,⁵ figure 4) untenable. Direct coil or liquid embolization of the venous varix itself was considered, but was unpalatable as the patient's symptoms related to mass effect on the brainstem, a condition that would not be ameliorated via this approach. Flow diversion was

Figure 3 (A) Lateral subtracted and (B) magnified lateral native fluoroscopy while the microvascular plug (proximal and distal markers at the bold and normal arrow) is deployed but not yet detached, demonstrating contrast stagnation in the dependent aspect of the venous varix (arrowhead). (C, D) Lateral projections of the right and left internal carotid artery injections, with the Micro Vascular Plug deployed, demonstrated a robust collateral supply to each posterior cerebral artery candelabra from each respective posterior communicating artery. There is also evidence of distal basilar artery (C, arrow) filling from a retrograde direction via the right posterior communicating and right P1. There is non-contiguous arteriovenous shunting (D, arrow) via the left P2, distinct from the venous varix of the basilar arteriovenous fistula (not visible in D).

Figure 4 (A) Immediate postoperative MRI axial T2 sequence demonstrating immediately decreased volume and decreased mass effect of the venous varix (asterisk) with lesional signal demonstrating early thrombosis (compared with [figure 1A](#)). (B) Lateral projection of MR angiogram from immediate postoperative MR angiogram reconstruction demonstrating internal carotid artery supply to the posterior cerebral artery via the posterior communicating artery bilaterally. The basilar artery terminates at the level of the anterior inferior cerebellar arteries. The varix is not seen. (C) Two-week follow-up MRI axial T1 sequence demonstrating thrombosis within the venous varix and diminution in mass effect compared with immediately after embolization (A). (D) Two-week follow-up MRI midline sagittal T1 sequence demonstrating thrombosis within the venous varix and diminution in mass effect compared with pre-embolization ([figure 1B](#)). (E) Three-month follow-up digital subtraction angiography with injection of the left vertebral artery demonstrates absent filling of the venous varix and upper basilar artery. (F) Lateral projection of the left vertebral artery angiogram at 3-month follow-up showing no opacification of the arteriovenous fistula.

considered but, in the setting of a high-flow fistula, this approach has achieved mixed success due to a persistent gradient across the stent wall. Moreover, given the patient's rapid and accelerating clinical decline, a treatment that would immediately decompress the varix was felt to be critical. Given the excellent posterior communicating artery collateral supply to the posterior circulation and basilar apex, focus shifted to sacrifice of the upper mid-basilar artery. Coil embolization of the basilar artery was considered, but the rapid antegrade flow into the AVF was such that all coil loops deployed instantly unfurled into the venous varix; moreover, adequate flow interruption with coils was unlikely without sacrificing a long segment of the basilar artery, including numerous non-collateralized perforators. Liquid embolic agents were also considered but, in the setting of high-flow arteriovenous shunting in the vicinity of critical perforators, inability to absolutely control the flow of a liquid risked devastating off-target embolization. Others have described transvenous embolization of cerebral arteriovenous malformations/fistulas and vein of Galen malformations quite dissimilar from this case.⁷⁻⁹ However, the concerns about control of liquid embolic agents and about mass effect on the brainstem precluded an analogous approach here.

Case series and literature reviews have found that advances in endovascular technology have improved treatment outcomes for pediatric AVFs, but there continues to be a high (60%) reported complication rate, necessitating a tailored approach for each lesion.³⁻⁶ Our application of a newer endovascular device represents a continued improvement in the available tools for treatment of these complex and often morbid lesions. The MVP device was proposed as a potential high-flow short-segment embolic device which was also reversible/retrievable during the procedure. Moreover, the structure of the device, with its open distal end posing no obstruction to continued flow to the basilar apex and its perforators, was particularly favorable in this location.

Positioning and deployment of the MVP device via a Hi-Flow Renegade 0.027 inch microcatheter (Marlborough, Massachusetts, USA) was uncomplicated, given the short throw of the deployment technique. Fortunately, the first deployment positioned the device with a polytetrafluoroethylene sidewall apposed directly to the fistulous point. Had the fistulous channel been wider than the sidewall of the MVP, more than one device may have been needed. Given the massive sump effect, we speculated that the fistulous basilar segment was unlikely to harbor functioning perforators and, fortunately, the sidewall did not induce brainstem ischemia via perforator

occlusion (both on clinical grounds and based on the lack of MRI diffusion restriction).

The remarkable improvement in symptoms suggested it was the fistulous flow into the varix and/or the mass effect that underlay the clinical symptomatology. The patient continues to undergo further evaluation of the risks of persistent arteriovenous shunting, particularly that of hemorrhage.

Contributors APS made substantial contributions to the conception and design of the work; acquisition, analysis, and interpretation of data for the work; drafting and critically revising the work for important intellectual content; final approval of the version to be published; and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. MAK made substantial contributions to the design of the work and interpretation of data for the work; drafting the work; final approval of the version to be published; and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. DBO made substantial contributions to the conception and design of the work and analysis and interpretation of data for the work; critically revising the work for important intellectual content; final approval of the version to be published; and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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