CASE REPORT

Retreatment of a choroidal vein of Galen malformation with embolization 42 years after open surgical treatment in the neonatal period

Lucas Eljovich,¹ Asim F Choudhri,² Adam S Arthur,¹ Paul Klimo,¹ Frederick A Boop,¹ Alejandro Berenstein³

¹Semmes-Murphey Clinic, Memphis, Tennessee, USA

²Department of Radiology, University of Tennessee Health Science Center, Memphis, Tennessee, USA

³Department of Neurosurgery, Icahn School of Medicine at Mount Sinai, New York, New York, USA,

Correspondence to

Dr L Eljovich, Semmes-Murphey Clinic, 6325 Humphreys Blvd, Memphis, TN 38120, USA; lejovich@semmes-murphey.com

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SUMMARY

In 1976 an infant boy initially presented with a choroidal vein of Galen malformation (cVOGM). Craniotomy and clipping of the anterior cerebral artery and posterior choroidal arteries was performed, which was reported in the literature 40 years ago. The patient remained asymptomatic until age 42 when he re-presented with an isolated intraventricular hemorrhage. Angiography demonstrated cVOGM with venous sinus occlusive disease leading to venous hypertension and subsequent intraventricular hemorrhage. The angiogram also demonstrated a 'pseudo-nidus' composed of multiple arterial-to-arterial anastomoses that had developed as a result of the original surgical treatment. We embolized a portion of the lesion to reduce the venous hypertension. The patient has been asymptomatic for more than 2 years. To our knowledge, this is the first reported case of a symptomatic neonatal VOGM treated with open surgery that required embolization as an adult for a delayed hemorrhagic presentation.

BACKGROUND

Vein of Galen malformation (VOGM) is a rare neurovascular disorder occurring in approximately one in every 25 000 live births.¹ The two angioarchitectural subtypes of VOGM—mural (mVOGM) and choroidal (cVOGM)—can have similar clinical presentations, including high output cardiac failure, hydrovenous disorders resulting in communicating hydrocephalus, developmental delay, seizures and (rarely) intracranial hemorrhage.² cVOGMs are more likely to present with high output congestive heart failure and require emergency treatment in the neonatal period due to the higher number of fistulous connections associated with it and the subsequent increased shunting.³

Prior to the development of modern neuroendovascular techniques, the morbidity and mortality of surgically treated VOGM was exceptionally high.⁴ Few children survived to adulthood with these lesions and little is known about the neurologic outcomes of the rare survivor of open surgery. Modern series of endovascular treatment of VOGM demonstrate that the majority of children can be treated successfully, with reported rates of angiographic cure of approximately 55% and—more importantly—much improved rates of normal neurologic outcome, ranging from 66.7% to 74% in the highest volume centers with extensive experience.^{3 5}

In this report we present the case of an adult who initially presented in the neonatal period with high output cardiac failure and then underwent open neurosurgical clipping of a portion of a VOGM. This treatment was reported in the literature 40 years ago.⁶ He subsequently re-presented 42 years later with an isolated intraventricular hemorrhage. To our knowledge, this is the first reported case of a symptomatic neonatal VOGM treated with open surgery that required embolization as an adult for a delayed hemorrhagic presentation. We describe his treatment course and discuss the unique angioarchitectural changes seen in the lesion as a result of the initial surgery and its impact on the endovascular treatment applied as an adult.

CASE PRESENTATION

A full term infant boy was born at 3.4 kg and developed failure to thrive and congestive heart failure by day 2 of life. Suspicion of an intracranial vascular malformation was confirmed by angiography that demonstrated a cVOGM.

Emergency surgery was undertaken and craniotomy and clipping of the anterior cerebral and posterior choroidal arteries was performed on day 8 of life. Due to persistent heart failure symptoms, a second surgery was performed 19 days later with clipping of an additional left posterior choroidal feeder. Although residual malformation was noted on follow-up angiography, the two surgeries resulted in resolution of the heart failure. The child suffered a perioperative stroke and developed epilepsy along with substantial motor and cognitive developmental delays. He was able to complete high school in a modified curriculum and is able to function independently as an adult despite a spastic hemiparesis of the left face, arm and leg.⁶

The patient remained asymptomatic until the age of 42 when he suffered a sudden severe headache and seizure. He was transferred to our center after a CT scan demonstrated intraventricular hemorrhage and a midline lesion concerning for a vascular malformation. MRI was not performed due to the presence of surgical clips predating MRI compatibility. The patient's neurologic examination was stable and diagnostic angiography was performed, which demonstrated a cVOGM with venous sinus occlusive disease (thrombosed right transverse and sigmoid sinuses), leading to venous hypertension and subsequent intraventricular hemorrhage. The angiogram also demonstrated a new component to the lesion—a

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'pseudo-nidus' composed of multiple arterial-to-arterial anastomoses proximal to the fistulous point in the region of the quadrigeminal cistern that was not present on the original angiogram (figure 1). This new component was thought to be due to neovascularization rather than recanalization of the clipped portion or residual malformation. We suspect that the placement of clips on the feeding arteries proximal to the exact junction with the venous sac in conjunction with the residual high flow lesion stimulated the recruitment of subependymal and choroidal arterial vessels.

TREATMENT

Due to the 'pseudo-nidus,' a direct path to the junction with the median vein of the prosencephalon was not possible. We therefore felt that it would be difficult to perform an embolization with N-butyl cyanoacrylate (n-BCA) and reach the fistulous point. A retrograde transvenous approach to treatment was deemed unfavorable due to the high risk of periprocedural hemorrhage associated with attempts at complete closure of the lesion via this retrograde technique.⁷ The goal of our initial treatment was not complete angiographic obliteration, but rather to attempt to balance the arterial inflow and venous outflow of the lesion in order to control the venous hypertension.

We chose to use Onyx 18 in conjunction with a Scepter balloon and Apollo detachable tip microcatheter in two separate left posterior cerebral artery pedicles. The advantage of the Scepter balloon is its ability to achieve immediate flow control without substantial reflux of embolic agent with balloon inflation. This allows Onyx to be pushed over a long distance through the network of arterial-to-arterial anastomoses to the point of the fistula.⁸ However, due to the higher profile (2.1 Fr) of the balloon catheter, its application is limited in extremely tortuous distal microvasculature. In contrast, the Apollo flow-directed 1.2 Fr microcatheter has a lower profile and is able to

gain more distal access in the microvasculature. Additionally, the detachable tip provides safety with regard to secure removal, despite the substantial embolic reflux that is often necessary when using the 'plug and push' technique. Using both strategies, we were able to embolize a substantial portion of the lesion despite the inability to navigate close to the site of fistualization due to the intervening arterial-to-arterial anastomosis that had developed as a result of the surgical treatment (figure 2).

OUTCOME AND FOLLOW-UP

The patient was seen in follow-up in the clinic and had experienced no new neurologic symptoms. We recommended continued transarterial embolization to exclude the lesion completely, but the family declined further treatment and he has been asymptomatic for more than 2 years.

DISCUSSION

VOGM can now be treated successfully with endovascular techniques, resulting in normal neurologic development. Disconnection of the precise arteriovenous fistulous point can result in symptomatic control and permanent closure of the lesion.³⁻⁹ We feel that n-BCA is the ideal embolic agent in most circumstances. It has multiple advantages over Onyx, including customizable concentration, its adhesive nature and rapid polymerization rate. However, in this case we chose to use Onyx in conjunction with a single-lumen balloon catheter and detachable tip catheters because of the angioarchitectural changes seen in the VOGM after proximal clipping of the feeding vessels.

Adult presentations of VOGM have been reported by multiple groups with a variety of symptoms including headaches, incidental radiologic findings, seizures, spontaneous thrombosis, coma and hemorrhage.¹⁰ The location of hemorrhage in the ventricular system is consistent with the choroidal localization of the lesion

Figure 1 Diagnostic cerebral angiography. Artist's representation (A) of the lateral unsubtracted vertebral angiogram (B) after the second craniotomy and clipping; there is no arterial network ('pseudo-nidus') seen below the venous collector (reproduced with permission from *JAMA Pediatrics*). Non-contrast head CT (C) demonstrates intraventricular hemorrhage from the malformation. Right internal carotid artery digital subtraction angiogram (D and E) demonstrates choroidal type vein of Galen malformation fed by multiple dilated posterior cerebral artery branches. There is no filling of the pericallosal artery with V-shaped surgical clip (black arrow) in the distal territory of the artery adjacent to the venous collector. There is occlusion of the right transverse sinus consistent with high flow venopathy. Left internal carotid artery digital subtraction angiogram (F and G) demonstrates similar findings to the right internal carotid artery injection with occlusion of right transverse and sigmoid sinuses (black arrow). Right vertebral artery injection (H and I) demonstrates enlarged vertebral and basilar artery consistent with longstanding high flow with multiple dilated thalamoperforators and posterior cerebral artery branches filling an arterial network of anastomosis—'pseudo-nidus' (black arrows) that was not present on the angiogram in the neonatal period after both surgeries.

Figure 2 Onyx embolization of a vein of Galen malformation. Anteroposterior and lateral high magnification left internal carotid artery injection (A and B) demonstrates a network of arterial-to-arterial anastomoses that prevent distal catheterization to the exact point of the fistula. Native image of Onyx 18 cast from the first pedicle (C and D) injected using an Apollo microcatheter (after removal, without distal tip detachment); a Scepter balloon microcatheter is in place in the second pedicle (pre-inflation). Left internal carotid unsubtracted angiogram high magnification (E and F) demonstrates the Onyx 18 cast in both pedicles and residual pseudo-nidus (black arrow in F). Left internal carotid artery injection post embolization (G and H) demonstrates devascularization of the two embolized pedicles with residual pseudo-nidus (black arrow in H).

and—in this specific case—correlates with the area of neovascularization with the subsequent ‘pseudo-nidus’ stimulated by proximal clipping of the feeding vessels during the neonatal period.

To the best of our knowledge, this case is the first report of a child who had open surgery in the neonatal period for VOGM and then required endovascular treatment as an adult for a subsequent hemorrhagic presentation. This case is also remarkable considering the four decades of clinical stability between initial symptoms and subsequent representation. This case emphasizes our incomplete understanding of the natural history of partially-treated VOGMs. Prior to the pioneering work and development of modern endovascular techniques by Lasjaunias and colleagues, normal neurologic survival was rare. These successes have led to the survival of a group of children who should be longitudinally followed. Given the modest rate of complete angiographic obliteration with modern techniques, it should be expected that the patient described in our report will not be unique with regard to delayed recurrent symptoms.

Learning points

- Prior to the development of modern neuroendovascular techniques, the morbidity and mortality of surgically treated vein of Galen malformation (VOGM) was exceptionally high.
- Neurologic outcomes with modern neuroendovascular techniques are favorable in a majority of patients.
- The long-term outcome of patients into adulthood with incompletely obliterated VOGM in childhood is incompletely understood. These patients require lifelong follow-up due to the risk of delayed symptoms including hemorrhage, as demonstrated by this unique patient.

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