Definitive treatment of vein of Galen aneurysmal malformation with stereotactic radiosurgery

Report of 2 cases

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Vein of Galen aneurysmal malformations (VGAMs) are uncommon congenital malformations arising from fistulous communication with the median vein of the prosencephalon, a primitive precursor of midline cerebral venous structures. Angiographic embolization is the primary modality for treatment given historically poor microsurgical outcomes. Only a few reports of treatment by Gamma Knife radiosurgery (GKRS) exist in the literature, and the results are variable. The authors present 2 cases of VGAM in which GKRS provided definitive treatment with good outcome: one case involving antenatal presentation of a high-output, mural-type VGAM with complex clinical course refractory to multiple embolic procedures, and the other a choroidal-type VGAM presenting with hemorrhage in an adult and without a feasible embolic approach. With discussion of these cases and review of the literature, the authors advocate inclusion of GKRS as a therapeutic option for treatment of these complex lesions.

Key Words • vein of Galen malformation • arteriovenous malformation • Gamma Knife radiosurgery • embolization • congenital malformation • neurosurgery • vascular disorders • stereotactic radiosurgery

Vein of Galen aneurysmal malformations are rare congenital arteriovenous anomalies occurring with an estimated frequency of less than 1 case per year at a neuroscience center serving a population of 3 million.2 In practice, they are often detected on prenatal ultrasound or are seen in the neonatal period in cases of high-output heart failure, with hydrocephalus, macrocephaly, and developmental delay characteristic of infant and early childhood presentation.7 Adult presentation is rare.14 The VGAM does not involve the developed vein of Galen but, rather, reflects arteriovenous shunting through its persistent embryological precursor, the median vein of the prosencephalon.11 The classic model excludes communication with deep venous drainage, and parenchymal cerebral venous flow is instead diverted through perimesencephalic and other collaterals,7 although exceptions to this rule have been reported.3,5,8

Abbreviations used in this paper: AVM = arteriovenous malformation; GKRS = Gamma Knife radiosurgery; ICA = internal carotid artery; PCA = posterior cerebral artery; VA = vertebral artery; VGAM = vein of Galen aneurysmal malformation.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
Radiosurgery for vein of Galen aneurysmal malformation

Case Reports

Case 1

This male patient prenatally diagnosed with a VGAM, without fetalis hydrops in utero, presented with high-output heart failure and respiratory distress upon delivery after approximately 35 weeks' gestation. His heart failure was initially managed medically until it necessitated intervention when the patient was 2.5 months of age, at which point the malformation had increased in size and the patient had developed mild hydrocephalus (Fig. 1A). Selective angiography of the right VA revealed a mural-type VGAM supplied by right posterior cerebral and superior cerebellar arterial feeders (Fig. 1B and C), which were subsequently coil embolized with good result, with complete angiographic occlusion of the VGAM (Fig. 1D) and improvement in hydrocephalus. Unfortunately, on follow-up imaging when the patient was 22 months of age, it was evident that the malformation had recanalized and had an appearance more of a choroidal-type VGAM, with the evolution of numerous collateral feeders arising from the anterior and posterior circulation. Angiography revealed a residual rete of vessels, below the vein of Galen sac, primarily fed by perimesencephalic and circumflex branches of the left PCA (Fig. 1E) and by a marginal tentorial branch from the left ICA (Fig. 1F), with disorganized medullary venous drainage and retrograde flow of the superior sagittal sinus and ophthalmic veins consistent with venous hypertension. The patient subsequently underwent a 2-stage embolization procedure involving transarterial cyanoacrylate injections of the anterior and posterior circulation, followed a week later by venous coiling of the residual vein of Galen sac and proximal straight sinus with restoration of antegrade venous flow. Persistent enlarged dural and posterior cerebral feeders were notable on MRI several months following embolization.

At 28 months of age, the patient presented to the emergency department after experiencing headache and generalized seizure; intraventricular hemorrhage originating from a new left thalamic venous varix was noted (Fig. 2A and B). Angiography revealed residual opacification along the anteroinferior aspect of the AVM. While general venous drainage from the head appeared to have stabilized compared to the prior angiogram, with distended superficial veins draining freely via the petrosal sinuses, we observed new anterior venous drainage through distended deep intrathalamic veins, including the ruptured left thalamic varix. An attempt at intervention was made by injecting cyanoacrylate into prominent posterior choroidal and right PCA feeders; however, a prominent medial thalamic perforator was not treated due to its critical nature. Postprocedure right ICA angiography demonstrated

![Image](https://example.com/image1)

**Fig. 1.** Case 1. A: Noncontrast CT scan of the head showing a dilated galenic sac (arrowhead) and mild hydrocephalus. B and C: Anteroposterior and lateral views of selective angiograms of the left VA showing a mural-type VGAM with enlarged galenic sac and torcula, with right superior cerebellar artery and PCA supply. D: Basilar artery angiogram following coil embolization of the arterial supply and galenic sac, confirming complete occlusion. E: Lateral selective angiogram of the left VA obtained at 22-month follow-up, revealing recurrence of a shunt, patency of the galenic sac, and transformation to a choroidal-type VGAM, with recruitment of perimesencephalic and circumflex branches primarily of the left PCA feeding a rete of vessels below the sac (arrow), with numerous new dilated venous collaterals also apparent (arrowhead). F: Additional contribution from the anterior circulation arises via a marginal tentorial branch of the left ICA (closed arrowhead), with a prominent posterior communicating artery (open arrowhead). Subsequent embolic intervention succeeded in restoring antegrade venous flow, without successful occlusion of the malformation.
diminished but persistent flow, with persistent opacification of the thalamic varix. Given the likelihood of rebleeding from the persistent varix and the residual malformation, inability to pursue further embolization, and absence of surgical options, the patient’s parents opted for their son to undergo GKRS. Preradiosurgery stereotactic angiography with GKRS fiducial markers demonstrated the extent of the residual feeding arteries and the patency of the problematic thalamic varix (Fig. 2C). When the patient was 31 months old, he underwent GKRS; a prescription dose of 17 Gy at the 50% isodose surface was delivered to the margins of the residual nidus (50% treatment volume = 10.9 cm³) using 33 isocenters, and no complications occurred (Fig. 2D). Subsequent follow-up MRI confirmed stability of the malformation without further development of anomalous arterial supply or venous drainage; involution of the thalamic varix was evident as early as 6 months postprocedure. At 18-month follow-up (Fig. 2E), studies showed thrombosis of the residual varix with a small amount of adjacent enhancement, and this was stable in configuration across several years of follow-up imaging. On last assessment, 6.5 years after hemorrhage and his subsequent definitive treatment, the patient has not experienced further sequelae of the malformation and is performing well in elementary school and sports, without discernable neurological deficit.

Case 2

This 27-year-old man with a history of AVM since birth presented with right thalamic hemorrhage. Magnetic resonance imaging demonstrated an enlarged galenic sac along with ventriculomegaly (Fig. 3A). Angiography revealed a large choroidal-type VGAM, with the nidus centered over the quadrigeminal cistern and draining, jet-like, into an enlarged galenic sac with a persistent falci-ne vein (Fig. 3B). Feeders included numerous enlarged left-sided circumflex mesencephalic and thalamoperforator arteries arising from the basilar artery and left PCA, along with a posterior medial choroidal artery arising from a fetal-type right PCA (Fig. 3C). The nature of the lesion precluded embolization as a treatment modality, and GKRS was chosen. The VGAM nidus received a radiosurgical prescription dose of 18 Gy at the 50% isodose surface using 17 isocenters (50% treatment volume = 6.4 cm³) (Fig. 4), and no acute sequelae occurred. The postoperative course was notable for a single seizure at 10 months; the patient was treated with Lamictal and there
was no seizure recurrence. Follow-up MRI at that time revealed a decreased AVM size, with thrombosis of the galenic sac (Fig. 3D). At the last follow-up 2.5 years after the radiosurgical procedure, the patient was without complaint, and his examination was notable for slight left-sided dysmetria on finger-to-nose test. Angiography showed complete resolution of the VGAM (Fig. 3E and F), with focal stenosis of the proximal left PCA and mild irregularity of adjacent smaller vessels of the posterior circulation (Fig. 3E), thought to correspond to sequelae of radiation arteritis.

Discussion

Two distinct cases of VGAM not amenable to conventional angiographic or open surgical treatment are presented in this report. The absence of other attractive management options, the high likelihood of progressive neurological disability in the absence of successful treatment, and the successful outcomes in these 2 patients lead us to advocate inclusion of GKRS as a therapeutic option in the treatment algorithm of these complex lesions.

The perinatal presentation of a mural-type VGAM in a patient with heart failure is somewhat atypical; choroidal lesions tend to present earlier with high-output failure.4 More notable is the dynamic nature of this lesion, with its transformation into a choroidal-type malformation over the course of treatment, characterized by the recruitment of neighboring choroidal arteries. The perinatal presentation of a mural-type VGAM in a patient with heart failure is somewhat atypical; choroidal lesions tend to present earlier with high-output failure.4 More notable is the dynamic nature of this lesion, with its transformation into a choroidal-type malformation over the course of treatment, characterized by the recruitment of neighboring choroidal arteries.

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of additional arterial contribution from the anterior and posterior circulation and the development of a nidus ventral to the recanalized galenic sac. With regard to the patient in Case 2, fewer than 20 cases of adult presentation of VGAMs have been previously reported. In both of our cases, GKRS provided definitive treatment in the absence of viable alternatives.

Previous reports illustrating the application of GKRS to VGAMs are sparse; no additional data have been published since 2000. In 1995 Tomsick et al. reported a case of choroidal VGAM in a 25-year-old woman with worsening headaches. A thalamoperforate/mesencephalic nidus was treated with 22.5-Gy (80% isodose) GKRS to a single isocenter as part of a staged treatment plan combining cyanoacrylate glue embolization and radiosurgery; angiography confirmed occlusion of the nidus. A contemporaneous report by Watban et al. that year documented the unsuccessful primary radiosurgical treatment of 3 VGAMs in infants 8–10 months of age using dosages of 20–25 Gy (50% isodose); all 3 patients initially presented with hydrovenous sequelae, without evidence of heart failure. Subsequent curative endovascular intervention was conducted when the patients ranged in age from 23 to 42 months, with one case suggesting development of mild retardation in the interim. The authors concluded that radiosurgery as a primary treatment modality for VGAMs was not indicated when an endovascular approach was feasible.

The most recent report published in 2000 by Payne et al., a case series of 9 patients who underwent radiosurgical treatment of malformations involving the vein of Galen, included 6 cases of true VGAMs by criteria of Lasjaunias and Raybaud. Three of these cases involved childhood presentation of mural VGAMs: 1 cure by GKRS at 6 years of age following failed embolization; 1 cure by alternating GKRS and embolization beginning when the patient was 11 years of age; and 1 with documented partial response to GKRS at 7 years of age following 3 failed embolizations, pending repeat GKRS at time of publication. Of the 3 choroidal cases, a cure was achieved in one patient with single primary GKRS therapy at 4 years of age, and in another patient shunting resolved with primary GKRS therapy at 6 years of age, with retreatment of subsequently revealed diminutive pericallosal feeders performed proximal to the time of that publication. In the third case, refractory to 4 sessions of embolization and with significant cognitive deficit, the patient had not had a response to GKRS therapy at 8 years of age. Doses ranged from 17 to 25 Gy (50% isodose). Of these 6 cases, clinically silent, radiation-induced change was noted in 1 patient, and there were no clinically apparent sequelae in the other 5 patients in whom a response to therapy was demonstrated.

Our 2 cases represent the youngest example of successful GKRS treatment of a VGAM and the only known case of definitive primary radiosurgery of a VGAM in an adult. While time to response is a significant criticism of GKRS therapy in the context of VGAMs, in our first case there was evidence of therapeutic response on 6-month follow-up studies and on 10-month studies in the second case. Outcomes were deemed satisfactory if not excellent, with normal childhood development in the first case and only slight dysmetria and sparse changes of radiation arteritis evident in the adult.

In clinically stable patients with lesions that preclude an endovascular embolic approach or are refractory to such therapy, our cases provide evidence that GKRS expands the therapeutic toolkit available for the treatment of VGAMs in adults and children.

Conclusions

The authors reported 2 cases, 1 perinatal and 1 adult, of VGAM in which GKRS provided definitive treatment with good outcome. After discussion of these cases and review of the literature, the authors advocate inclusion of GKRS as a therapeutic option for treatment of these complex lesions.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Triffo, Morris. Acquisition of data: Couture, McMullen, Tatter, Morris. Analysis and interpretation of data: Triffo, Bourland, McMullen, Tatter, Morris. Drafting the article: Triffo, Bourland. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Triffo.

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Accepted June 6, 2013.

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Please include this information when citing this paper: published online July 19, 2013; DOI: 10.3171/2013.6.JNS121897.

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