Selection of Endovascular Approach of Vein of Galen Aneurysmal Malformation

O. ITO^{1,2}, K. GOTO³, N. OGATA¹, H. UTSUNOMIYA⁴, S. SATO³, A. FUKUMURA⁵

Department of Interventional Neuroradiology; Iizuka Hospital¹

Department of Neurosurgery; Shin-Koga Hospital²

Brain Attack Center; Ohta Memorial Hospital³

Department of Radiology; Fukuoka University⁴

Department of Neurosurgery; Shimonoseki Kousei Hospital⁵

Key words: vein of Galen aneurysmal malformation, endovascular treatment, transarterial embolization, transvenous embolization

Summary

Vein of Galen aneurysmal malformation (VGAM) is one of the most difficult intracranial vascular lesions because this disease consists of extremely high flow shunts and affects infants and small children. Thanks to the development of various diagnostic modalities, early diagnosis became possible allowing us to prepare appropriately according to the patients' general and neurological conditions. Recent improvements of endovascular techniques and materials enabled both transarterial and transvenous approaches even to the newborn infants, widening therapeutic windows. In this article, we discuss the selection of endovascular approaches based upon angioarchitecture of VGAM presenting four representative cases from our file.

Introduction

VGAM is a rare anomaly which affects the pediatric population. This disorder represents less than 1% of all the intracranial arteriovenous malformations (AVM's). The natural history of the disease is haunted by high mortality and morbidity rates¹. VGAM is an arteriovenous fistula occurring at the end of the embryonic period. It represents a dilatation of the persistent embryonic median prosencephalic vein of

Markowski, with direct arteriovenous shunts². Lasjaunias and colleagues have proposed that VGAMs be subclassified into two distinct types: choroidal and mural type³. In the choroidal type, multiple fistulas are located in the cistern of the velum interpositum anywhere within the tela choroidea. The shunt is extracerebral, subarachnoid, and communicates with the anterior aspect of the median vein of the prosencephalon.

The arterial feeders are usually bilateral and correspond to the normal choroidal arteries, subfornical branch or pericallosal artery, or subependymal branches of thalamoperforators. This type of VGAM usually causes heart failure in newborns and can be recognized by its complex arterial network.

In the mural type, the fistula is in the wall of the median prosencephalic vein, usually in its inferolateral margin and is also extracerebral and subarachnoid. The vessels supplying the shunt are usually the collicular and/or the posterior choroidal arteries and may be unilateral or bilateral. Clinically, the mural types of VGAM present later in infants with macrocephaly or failure to thrive and may be associated with mild cardiac failure or asymptomatic cardiomegaly.

A review of the literature, regarding to the

Figure 1 Case 1 a) CT scan at 14 months of age showing aneurysmal dilatation of the vein of Galen (asterisk) and hydrocephalus. b) Vertebral angiogram, lateral view, showing posterior choroidal arteries as feeding pedicles (arrowhead) and prosencephalic vein as draining vein (arrow). c) Venogram during transthoracic embolization showing coils in the vein of Galen (arrow).

Figure 2 Case 2 a,b) Prenatal MRI showing aneurysmal dilatation of the vein of Galen (asterisk). c) Vertebral angiogram, Towne projection, showing collicular and posterior choroidal arteries as feeding pedicles (arrows) and aneurysmal dilatation of the vein of Galen (asterisk). d) Compare the above mentioned legends with the MR angiogram which is non-invasive and useful in planning endovascular treatment. e) Deposition of NBCA after superselective cannulation of the posterior choroidal artery up to the fistulous point. f) T1-weighted image, sagittal view, at 11 days after transarterial embolization. Note VGAM entirely filled with thrombus without flow-void.

Figure 3 Case 3 a) T2-weighted image, axial view, showing aneurysmal dilatation of the vein of Galen (asterisk), macrocephaly and mild diffuse cerebral atrophy. b) T2-weighted image, sagittal view, showing feeders as anterior pericallosal arteries and drainers as prosencephalic vein (double asterisk). c,d) Deposition of NBCA after superselective cannulation into the bilateral posterior choroidal arteries. c) AP and d) lateral projection. Note GDCs deposited before injecting NBCA. e,f) Vertebral angiogram, lateral view, during transvenous embolization showing detachable coils in the venous pouch. e) Two microcatheters were placed simultaneously for the first session (arrow). f) One microcatheter was used during the second session. Note marked reduction in degree of dilatation of the numerous feeding pedicles and reduction of flow through the fistula.

natural history and results of surgical treatment, shows that the outcome has been singularly depressing. The natural history of VGAM patients is characterized by a marked psychomotor developmental delay and very high mortality rate^{1,3}. Results of conventional treatments are far from satisfactory¹. A major breakthrough in the treatment of patients with VGAMs began in the early 1980s with the use of endovascular embolization by transtocular approach. Encouraged by the successful results, less invasive transfemoral transarterial and transvenous embolization were developed with the advent of microcatheters^{3,4,5}. We present 4 representative cases from our file and discuss the timing of treatment and selection of endovascular approach based upon image diagnosis findings.

Case 1

This was the initial case treated by us in the mid-80's before the advent of microcatheters. After taking normal developmental milestone, this baby boy developed frequent seizures and became lethargic at 14 months of age. Brain CT showed a large Vein of Galen aneurysmal malformation and hydrocephalus (figure 1a). Angiography disclosed a mural-type VGAM supplied by enlarged posterior choroidal arteries bilaterally (figure 1b). After placing a burr hole on the external protuberance, the torcular of the sinuses was directly punctured and the vein of Galen was packed with multiple 0.035 Gianturco fibered coils (figure 1c). Approximately 100 coils were deposited in 3 separate sessions and marked flow reduction was attained. Complication of this procedure was moderate ventricular hemorrhage which was identified

Figure 4 Case 4 a) T1-weighted image, sagittal view, showing aneurysmal dilatation of the vein of Galen (asterisk). b) Internal carotid angiogram, lateral view, after embolization of the posterior choroidal arteries using GDCs and NBCA showing anterior pericallosal artery as a residual feeding pedicle. c) T1-weighted MRI, sagittal view, taken 5 months after embolization. Note marked decrease of VGAM with partial thrombosis.

by CT examination after the second session. The hemorrhage subsided quickly without needing any treatment. The patient showed temporal improvement of activity right after each session. However, the patient also had high fever and developed status epilepticus after each procedure that worsened the neurological status. The treatment was terminated because the patient's family did not want the continuation of the treatment.

Case 2

The VGAM was suspected in this baby girl by regular maternity check-up using a Doppler ultrasonography. The diagnosis was established at 30 weeks by prenatal MRI examination. It was also shown that there was no cerebral atrophy, hydrocephalus and other cerebral anomaly (figure 2a,b). The baby girl was born at 37 weeks by the Caesarian section. She was physically intact at birth and took normal developmental milestone thereafter. As the follow up MR showed slight ventricular dilatation at 8 months of age, she was referred to us for the treatment. On admission, the patient was slightly hypoactive and presented with dilated facial veins in the forehead. She was neurologically intact and did not show increased head circumference and cardiac symptoms. (figure 2d). Angiography revealed that the malformation was supplied by dilated bilateral posterior choroidal arteries and was classified to be a mural-type VGAM (figure 2c,d). The patient was treated by transarterial embolization in 2 sessions. A microcatheter was easily advanced deep into the dilated feeding arteries and the

fistulous sites were occluded with pure NBCA (N-butyl-2-cyanoacrylate) (figure 2e). MRI at 11 days after embolization showed complete thrombosis of the VGAM without any flow-void (figure 2f). Dilated facial veins disappeared and her activity level improved markedly since the day after the treatment. A follow-up MRI obtained 18 months later showed a marked shrinkage of the thrombosed VGAM and disappearance of ventricular dilatation. She welcomed her birthday healthily and had developmental landmark.

Case 3

This girl with mild psychomotor developmental delay had occasional headache and vomiting at 3 years of age. MRI performed at outside hospital revealed a VGAM, hydrocephalus and mild brain atrophy (figure 3a,b) and referred to us for the curative treatment. On admission, she presented with increased head circumference and dilated cranio-facial vein. No cardiac symptom was noted. Cerebral angiography demonstrated the VGAM to be a choroidal-type supplied by enlarged bilateral posterior choroidal arteries, thalamoperforating arteries and anterior pericallosal arteries. In this case, a combination of transarterial and transvenous embolizations was performed. In order to safely deposit NBCA to obliterate the extremely high flow feeding pedicles and the fistulous point, a few GDCs were placed prior to injection of NBCA (figure 3c,d). Transvenous embolization was performed in three sessions. Two microcatheters were navigated simultaneously into the venous pouch and multi-

ple detachable platinum coils, i.e. Galenic coils and IDCs, were placed and detached one at a time. Considerable numbers of fibered coils were intermittently weaved into the coil mesh to facilitate thrombus formation (figure 3e,f). Headache and nausea immediately disappeared after embolization. MRI at 3 months after embolization showed disappearance of flow of VGAM. A follow-up MRI obtained 3 years later showed MRI showed dilation of ventricular system and brain atrophy, despite complete closure of the fistula. The patient's growth and development have caught up to a normal level.

Case 4

This baby boy was born at 38 weeks gestational period. The Apgar score was 9 points at birth, but he sometimes presented with cyanosis which spontaneously regressed. At one month after birth he presented with apnea and cyanosis, and was diagnosed to have VGAM by CT and MRI (figure 4a). On admission, the patient showed no neurological deficit, but presented with mild cardiac failure. Cerebral angiography demonstrated a choroidal-type VGAM supplied by bilateral posterior choroidal arteries and pericallosal arteries (figure 4b). Because of the presence of multiple feeding pedicles, combined transarterial and transvenous embolization was performed. After depositing a few GDCs in the feeding pedicles, pure NBCA was deposited to obliterate main feeding pedicles and the fistulous point (figure 4b). Transvenous embolization was performed in 2 sessions a few weeks later. He no longer developed seizures immediately after embolization. Cardiac failure disappeared completely and his growth and development was normal. MRI at 5 months after embolization showed marked shrinkage of the VGAM which is largely thrombosed (figure 4c). Although a small part of the fistula persisted, treatment was discontinued because the patient's family did not want further treatment.

Discussion

Indication and timing of treatment

The patients with VGAM develop heart failure, hydrocephalus, neurological disorders, and multiple organ failure because of the extremely high flow arteriovenous shunt. Therefore, deci-

sion of the optimal timing for curative treatment is crucial for the prevention of irreversible changes of brain and other organs⁶. A neonate with congestive heart failure refractory to medical treatment may require emergent intervention. If neonatal examinations suggest such possibility, a medical team consisted of endovascular therapist, pediatric cardiologist, pediatric anesthetologist and neonatologist should stand-by delivery in order that necessary treatment could be instituted immediately.

Transtorcular approach

Transtorcular embolization following craniotomy had been the only possible approach for transvenous embolization before the development of microcatheters and microguidewires⁵. This approach allows quick delivery of large fibered coils, and more dramatic flow reduction can be achieved than the transfemoral or transjugular venous embolization using bare platinum coils. As arterial inflow persists during embolization, transvenous embolization should be staged into several sessions in order to prevent intracranial hemorrhage due to abrupt pressure elevation⁷. Other complications reported were vessel perforation during coil deposition, abscess formation in the coil mass deposited in the VGAM and wound infection.

Endovascular treatment

Development of endovascular treatment have achieved satisfactory morphological and/or clinical results⁸. Endovascular approaches include transarterial and transvenous embolization, often performed in combination and as multi-staged operation.

Transfemoral transarterial embolization

Superselective catheterization of intracranial feeders supplying arteriovenous fistulas is performed up to the fistulous point using a combination of a soft microcatheter and microguidewire. The feeder is then occluded with NBCA⁸. As this is an extremely high flow shunt, it is safer to place a few detachable coils before injecting NBCA for the accurate deposition of the glue in the fistulous coil. Mural type VGAM can be completely obliterated using transarterial embolization alone (Case 2).

Transfemoral transvenous embolization

Transveous approach is principally for the

embolization of the choroidal type VGAM supplied by multiple feeding pedicles. Also this approach is needed for cases with mural type VGAM in which transarterial embolization failed to completely obliterate fistulae. Sometimes we have to resort to transvenous embolization because of the excessively tortuous feeding pedicles that prevent superselective cannulation up to the fistulous point⁹. Selective catheterization of Galenic pouch via transfemoral transvenous approach is performed and then detachable platinum coils are placed carefully.

Selection of coil with optimal diameter is important for the prevention of coil migration. If we are not sure the diameter of the largest coil exceed that of the orifice of the venous exit, it may be useful to place two catheters simultaneously into the Galenic pouch and detach tangled coils alternatively. However, for the prevention of intracranial hemorrhage and venous infarction resulting from sudden increase in venous pressure, multi-staged embolization or combined transarterial and transvenous embolization are performed.

Usefulness of modern diagnostic modalities

Doppler sonography and MRI have contributed to prenatal diagnosis of VGAM¹⁰. Doppler sonography is possible to diagnose it prenatally, easily, and non-invasively. Prenatal MRI provides us of accurate information in-

cluding the status of brain parenchyma and ventricular system¹¹. Prenatal diagnosis by these modalities is important for the decision of indication of the treatment and therapeutic approach. Cerebral angiography is still the most useful and accurate diagnostic tool and is prerequisite for endovascular treatment, however, as it is invasive for infants, it should be performed in the same session as the endovascular treatment. MRA enabled us to understand angioanatomy non-invasively, and allowed us to design endovascular treatment beforehand. Also, MRI and MRA are useful tool for follow-up after treatment¹².

Conclusion

The patients with VGAM develop heart failure, hydrocephalus, neurological disorders, and multiple organ failure because of the extremely high flow arteriovenous shunt. Therefore, decision of the optimal timing for curative treatment is crucial for the prevention of irreversible changes of brain and other organs. Thanks to the development of various diagnostic modalities, however, early diagnosis became possible allowing us to prepare appropriately according to the patients' conditions, and the improvement of endovascular techniques and materials enabled us both transarterial and transvenous approach even to the newborn infants widening therapeutic windows.

References

- 1 Johnston IH, Whittle IR et Al: Vein of Galen malformation: diagnosis and management. *Neurosurgery* 20: 747-758, 1987.
- 2 Raybaud CA, Strother CM et Al: Aneurysms of the vein of Galen: embryonic considerations and anatomical features relating to the pathogenesis of the malformation. *Neuroradiology* 31: 109-128, 1989.
- 3 Berenstein A, Epstein F: Vein of Galen malformations: combined neurosurgical and neuroradiologic intervention, Arteriovenous fistulas of the brain. In: *Surgical Neuroangiography: vol 4. Endovascular Treatment of Cerebral Lesions*. Springer Verlag, Heidelberg 1992: 267-317.
- 4 Lasjaunias P, Rodesch G et Al: Treatment of vein of Galen aneurysmal malformation. *J Neurosurg* 70: 746-750, 1989.
- 5 Mickle JP, Quisling RG: The transtorcular embolization of vein of Galen aneurysms. *J Neurosurg* 64: 731-735, 1986.
- 6 deKoning TJ, Gooskens R et Al: Arteriovenous malformation of the vein of Galen in three neonates. *Eur J Pediatr* 156: 228, 1997.
- 7 Lylyk P, Vinuela F et Al: Therapeutic alternatives for vein of Galen vascular malformations. *J Neurosurg* 78: 438-445, 1993.
- 8 Lasjaunias P, Garcia MR et Al: Vein of Galen malformation. Endovascular management of 43 cases. *Childs Nerv Syst* 7: 360-367, 1991.
- 9 Casasco A, Lylyk P et Al: Percutaneous transvenous catheterization and embolization of vein of Galen aneurysms. *Neurosurgery* 28: 260-266, 1991.
- 10 Campi A, Scotti G et Al: Antenatal diagnosis of vein of Galen aneurysmal malformation: MR study of fetal brain and postnatal follow-up. *Neuroradiology* 38: 87, 1996.
- 11 Yuval Y, Lerner A et Al: Prenatal diagnosis of Vein of Galen aneurysmal malformation: report of two cases with proposal for prognostic indices. *Prenat Diagn* 17: 972, 1997.
- 12 Campi A, Rodesch G et Al: Aneurysmal malformation of the vein of Galen in three patients: clinical and radiological follow-up. *Neuroradiology* 40: 12, 1998.

Osamu Ito, M.D., Ph.D.
Department of Neurosurgery
Tenjin-Kai Shin-Koga Hospital
120 Tenjin-cho, Kurume, Japan 830-8577