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Vein of Galen Aneurysmal Malformation in a Neonate Treated by Endovascular Surgery

—Case Report—

Masaki KOMIYAMA, Misao NISHIKAWA, Toshihiro YASUI, Shouhei KITANO*, Hiroaki SAKAMOTO*, and Takeshi INOUE**

*Departments of Neurosurgery, *Pediatric Neurosurgery, and **Pathology, Osaka City General Hospital, Osaka*

Abstract

A male neonate with vein of Galen aneurysmal malformation developed fetal heart failure after a gestation period of 37 weeks. Labor was induced, but a cesarean section had to be performed because of fetal distress. His Apgar scores were 2/8 at birth. The boy required controlled ventilation due to generalized seizure and cyanosis. Neuroimaging revealed vein of Galen aneurysmal malformation with enormous arteriovenous shunt flows. Transarterial embolization was performed on days 4 and 6 to reduce the progressive high-output congestive heart failure. Three large feeders were occluded with interlocking detachable coils, markedly reducing the shunt flow. Although heart failure improved markedly, he died of pulmonary hemorrhage on day 12. Endovascular surgery can improve the prognosis of heart failure in the neonatal period less invasively than other treatment modalities, and may help to minimize ischemic brain damage and thus increase the possibility of normal brain maturation.

Key words: embolization, heart failure, neonate, vein of Galen, aneurysmal malformation

Introduction

Vein of Galen vascular abnormalities are extremely rare. There are several methods of classification,[2,5,7,9,13,21] but a simple and useful way is based on the presence or absence of an arteriovenous nidus. Vein of Galen aneurysmal malformation is caused by direct arteriovenous fistula, whereas vein of Galen aneurysmal dilatation is caused by arteriovenous malformation. This classification separates patients into two groups with different natural histories and different optimal treatment modality.

Vein of Galen aneurysmal malformation is a congenital arteriovenous fistula due to persistence of the median vein of the prosencephalon occurring between 6 and 11 weeks of development.[19] The abnormal vein does not drain normal cerebral tissue and thus does not communicate with the normal cerebral veins. This malformation is further classified as mural or choroidal according to the location of the fistula in the wall of the aneurysmal sac or in the cistern of the velum interpositum. Vein of Galen aneurysmal dilatation consists of the dilated true vein of Galen that drains normal cerebral tissue as well as abnormal shunted flow, and can be classified into parenchymal arteriovenous malformations, dural arteriovenous fistulas, and varices.[7]

Vein of Galen aneurysmal malformation in neonates usually results in progressive high-output congestive heart failure. Treatment is always challenging, and the prognosis is extremely poor.[10-12] Here we present a neonatal patient with vein of Galen aneurysmal malformation and discuss the management of vein of Galen aneurysmal malformation.

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Case Report

A neonatal boy was the second child of healthy parents. The first child was normal. Antenatal Doppler sonography at a gestation period of 32 weeks had disclosed an intracranial cystic vascular lesion. Follow-up sonography at 34 weeks revealed an enlarged heart (cardiothoracic area ratio of 40%) and enlarged intracranial lesion with a diameter of 19 mm

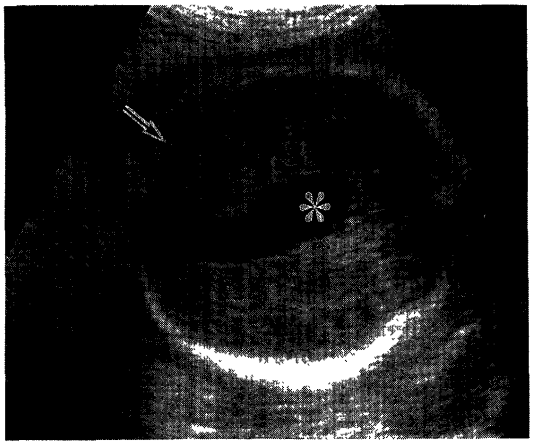

Fig. 1 Doppler sonogram at a gestation period of 34 weeks revealing a large cystic lesion 19 mm in diameter (*asterisk*) at the center of the head, corresponding to the aneurysmal sac of the vein of Galen aneurysmal malformation, and the transverse sinus (*arrow*).

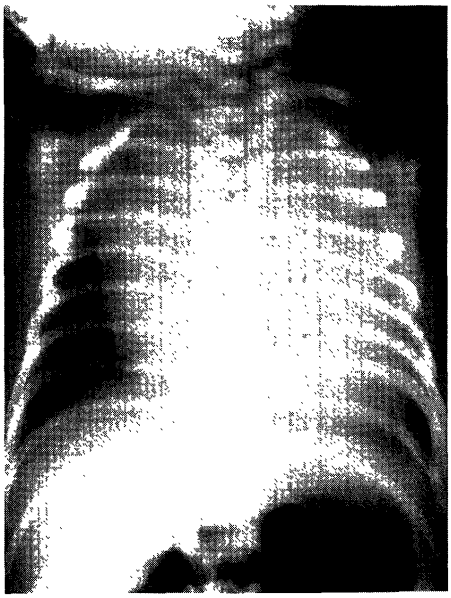

Fig. 2 Chest radiograph on admission (day 0) showing the enlarged heart with a cardiothoracic ratio of 79%. The right atrium and the right ventricle are especially enlarged.

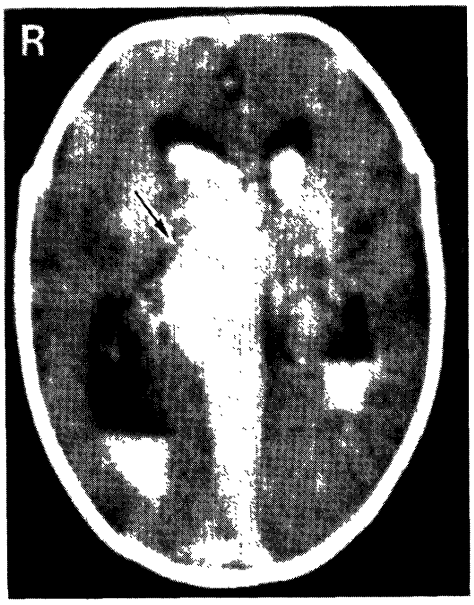

Fig. 3 CT scan revealing a slightly high density midline structure (*arrow*) and intraventricular hemorrhage.

(Fig. 1). Progressive fetal cardiac failure was found at 37 weeks (cardiothoracic area ratio of 42% and cardiothoracic ratio of 69%). Therefore, induction of labor was attempted but resulted in fetal distress, requiring an emergency cesarean section. At birth he had Apgar scores of 2/8, a birth weight of 2860 g, and a head circumference of 33.4 cm. The boy was transferred to our hospital 5 hours after birth for further evaluation and treatment.

On admission, he was inactive and obviously obtunded. No apparent motor weakness was observed. The fontanels intensely pulsated in synchrony with the heartbeat. No dilatation of the facial or scalp veins was observed. Loud cranial bruit was present everywhere over the head. He developed generalized seizure and cyanosis, requiring controlled ventilation under endotracheal intubation.

Skull radiography showed mild suture separation but no calcification. Chest radiography demonstrated marked cardiomegaly (cardiothoracic ratio of 79%) (Fig. 2). Doppler sonography revealed vein of Galen aneurysmal malformation, persistent ductus arteriosus (right to left shunt), persistent foramen ovale (right to left shunt), dilatation of the right atrium and ventricle, and pulmonary hypertension. Computed tomography (CT) showed intraventricular and subependymal hemorrhage and a slightly high density midline structure, which was homogeneously enhanced by the contrast material (Fig. 3). Magnetic resonance (MR) imaging and MR angiography revealed subependymal hemorrhage, multiple large feeding vessels, and a large aneurysmal sac approximately 18 mm in diameter draining to the large straight sinus (Fig. 4).

Under controlled ventilation, catecholamine was administered to improve cardiac failure from day 0. However, cardiac failure did not improve. Then administration of prostaglandine E_1 and nitric oxide in-

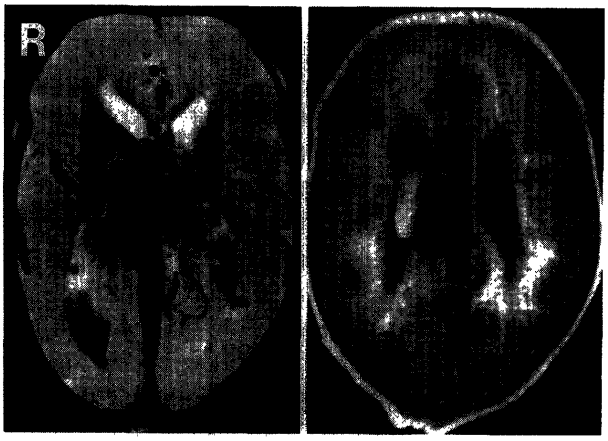

Fig. 4 *left*: T_1-weighted MR image revealing the aneurysmal sac and straight sinus as a signal void, and intraventricular hemorrhage. *right*: T_2-weighted MR image clearly showing the subependymal hemorrhage.

halation were started to reduce pulmonary resistance from day 2, which improved his cardiopulmonary status and the shunt flow in the ductus arteriosus changed from left to right.

On day 4, cerebral angiography was performed under general anesthesia using a biplanar digital subtraction angio-system (DFP-60A; Toshiba Medical Inc., Tokyo), which revealed a large vein of Galen aneurysmal malformation (choroidal type) fed by the bilateral pericallosal arteries of the anterior cerebral arteries, bilateral distal middle cerebral arteries, many feeders from the bilateral posterior cerebral arteries including the posterior choroidal arteries, and the left posterior inferior cerebellar artery. An anastomotic arterio-arterial maze was interposed between the feeders and the aneurysmal sac. The large aneurysmal sac was connected to the straight sinus and then to the torcular herophili. There was no evidence of reflux of the shunted flow to the deep cerebral veins. The normal venous flow draining the cerebral tissue was not visualized (Fig. 5A–D).

We started by performing diagnostic angiography with a 3 French femoral sheath and proceeded to transarterial coil embolization. The microcatheter (FasTracker-18 catheter; Target Therapeutics Inc., Fremont, Cal., U.S.A.) and microguidewire (0.014 inch) were difficult to navigate into the target vessels without a guiding catheter, because the microcatheter was unstable in the vertebrobasilar system and easily fell into the aorta. After maneuvering the catheter for about 2 hours, the 3 French sheath was exchanged for a 4 French sheath and a 4 French guiding catheter (Glidecath II, H1H; Terumo Inc., Tokyo). This 4 French system markedly facilitated the interventional procedures. Two large feeders, the left posterior inferior cerebellar artery and the right lateral posterior choroidal artery, were occluded

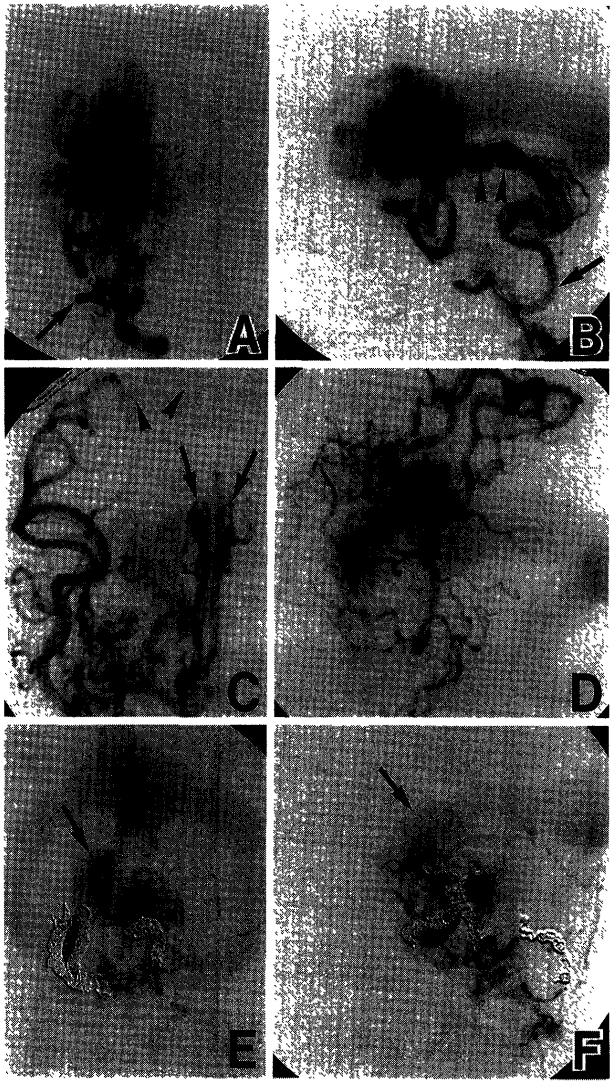

Fig. 5 Left vertebral angiograms, anteroposterior (A) and lateral views (B), showing the vein of Galen aneurysmal malformation. The left posterior inferior cerebellar artery (*arrow*) feeds the aneurysmal sac through an arterio-arterial maze (*arrowheads*). Right carotid angiograms, anteroposterior (C) and lateral views (D), showing the bilateral anterior cerebral arteries (*arrows*) and the right distal middle cerebral arteries (*arrowheads*) contributing to the arteriovenous shunt. Post-embolization left vertebral angiograms on day 6, anteroposterior (E) and lateral views (F), revealing marked reduction of the shunted flow. The diameter of the aneurysmal sac (*arrow*) has been reduced from 18 to 9 mm.

Fig. 6 Plain skull radiographs, anteroposterior (*left*) and lateral views (*right*), showing the coils in the embolized feeding arteries.

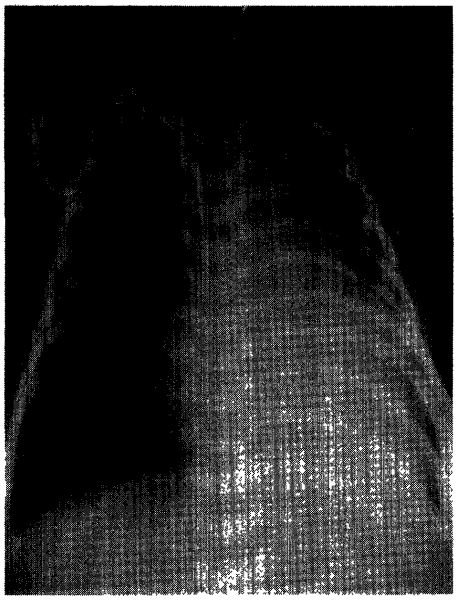

Fig. 7 Chest radiograph on day 6 showing markedly improved cardiac failure (cardiothoracic ratio of 62%).

with interlocking detachable coils (Target Therapeutics Inc.) in the proximal portion of the arterio-arterial maze of the fistula. A total dose of about 20 ml of iopamidol (300 mgI/ml) was used. The femoral sheath was left in place.

On day 6 a second intervention was carried out to further reduce the shunted flow. Another large feeder, the right medial posterior choroidal artery, was occluded in the same manner with interlocking detachable coils (Fig. 6). Final angiography revealed marked reduction of the shunted flow and reduced diameter of the aneurysmal sac from 18 to 9 mm (Fig. 5E, F). The total dose of contrast material was 12 ml.

His congestive heart failure improved markedly (cardiothoracic ratio of 62%), but his general condition gradually deteriorated, probably due to persistent pulmonary hypertension that was refractory to vigorous medical treatment (Fig. 7). On day 9 he developed a pulmonary hemorrhage, which caused respiratory failure and he died on day 12.

An autopsy was performed. The brain weighed 176 g and appeared markedly atrophic. Subarachnoid hemorrhage was noticeable, especially in the prepontine and basal cisterns. The aneurysmal sac was located at the center of the brain and had thick walls like the dural venous sinus (Fig. 8 *left*). The three embolized vessels were occluded with coils and thrombus. There was no perforation of the vessels by the coils, which could be seen through the thin, transparent vascular walls. The feeding arteries occluded with coils had a diameter of 3–4 mm, but the non-embolized feeders were completely collapsed and were easily severed during dissection (Fig. 8 *right*). There were many fine arterial mazes between the feeding arteries and the aneurysmal sac. The anterocaudal aspect of the aneurysmal sac was adherent to the surrounding structures, such as the bilateral thalami and the roof of the third ventricle.

The heart weighed 43 g. The right atrium and right ventricle were markedly dilated. The foramen ovale and ductus arteriosus were both patent. The lungs showed marked bilateral hemorrhaging and congestion. The left femoral artery in which the vascular sheath was placed was patent.

Discussion

I. Symptomatology

Choroidal type of vein of Galen aneurysmal malformation occurs mostly in neonates who present with congestive heart failure due to overload caused by the shunted flow. Mural type of vein of Galen aneurysmal malformation is predominantly observed in infants who present with hydrocephalus, macrocephaly, mild heart failure, or seizures. In older children, the main manifestations are focal neurological deficits, headache, and subarachnoid hemorrhage.[9] Hydrocephalus is attributable to compression of the aqueduct or abnormal cerebrospinal fluid turnover due to high venous pressure.[21]

II. Angioarchitecture

Feeding arteries may include the anterior and posterior choroidal arteries, the pericallosal arteries of the anterior cerebral arteries, the middle cerebral

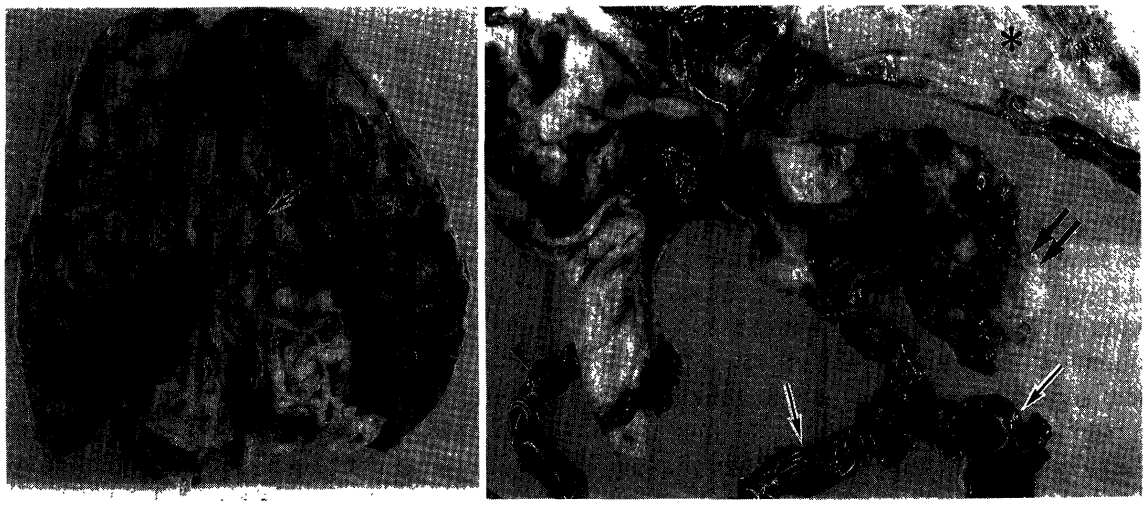

Fig. 8 *left*: Photograph showing the gross anatomy of the brain viewed from the back. The corpus callosum has been severed to obtain a better view of the aneurysmal sac (*arrow*). The falx cerebri is reflected backward. *right*: Photograph showing the specimen of the vein of Galen aneurysmal malformation with the falx cerebri (*asterisk*). The entry of many feeding vessels can be seen through the opening of the aneurysmal sac (*double arrow*). Interlocking detachable coils can be seen in the occluded left posterior inferior cerebellar artery (*arrowhead*) and the occluded posterior choroidal arteries (*arrows*).

arteries, and the anterior and posterior thalamoperforating arteries, or sometimes the meningeal and cerebellar arteries.[19] An anastomotic arterio-arterial maze is often interposed between the feeders and the fistula,[19] as in our patient. The falcine sinus often drains to the superior sagittal sinus instead of the occluded straight sinus. This venous route frequently displays stenosis, which reduces congestive heart failure but increases the size of the aneurysmal dilatation.

III. Diagnosis

Doppler sonography is the primary diagnostic tool because of its convenience at bedside, non-invasiveness, and real-time capability. Conventional cerebral angiography (digital subtraction angiography) is still the basis for diagnosis even though CT and MR imaging are less invasive. CT demonstrates a spherical posterior third ventricular mass due to the density of the circulating blood, which is homogeneously enhanced by the contrast material.[22] CT and MR imaging clearly show hydrocephalus and brain damage caused by ischemia or hemorrhage.[16] These modalities are especially useful in neonates because vascular access and the dosage of contrast material for angiography are strictly limited.[21] MR imaging can provide more information than CT about anatomical structure, including the vasculature and brain damage.

IV. Angiographic technique

Transfemoral angiography is possible using a 4 French catheter through the femoral vascular sheath. Leaving this sheath in place, repeated angiography and/or transarterial embolization can be performed over several days. The microcatheter can be navigated into the intracranial vessels through a diagnostic 4 French catheter. Biplanar digital subtraction angioequipment can reduce the time required for the diagnostic and/or interventional procedures as well as the amount of the contrast material necessary.

We considered the upper dose limit of contrast material to be 6 ml/kg weight.[8] Heparin in the saline solution for angiography should be minimized to avoid abnormal coagulation and possible hemorrhagic complication. We left the femoral sheath in place for 3 days. Microinfusion of heparinized saline kept the sheath patent and systemic heparinization was avoided. Autopsy revealed that the femoral artery was still patent, suggesting that systemic heparinization is unnecessary. It is extremely important to keep the body temperature normal during angiography and intervention. Decreased body temperature easily leads to deterioration of general condition in neonates. It is essential to use warmer blankets and to wrap the extremities with cotton and aluminum foil.

V. Treatment

Medical treatment is indicated to control congestive heart failure in neonates. It includes the use of di-

uretics, catecholamine, and controlled ventilation. To treat pulmonary hypertension, administration of prostaglandine E_1 and nitric oxide inhalation are indicated. Surgical treatment has been successful, but it is extremely difficult in neonates.[24] Therefore, endovascular treatment is increasingly being employed.[6,8,12,14] Radiotherapy offers little for neonatal patients because the time required to achieve a significant result is too long for the rapidly developing and maturing brain, leading to irreversible mental retardation.[1] If the patient's clinical status allows, treatment should be delayed to ensure the optimal timing for definitive treatment.[8] Angiography should also be delayed since CT and MR imaging can provide enough information and the femoral artery should be kept intact for future intervention. Embolization appears to be the best method to treat hydrocephalus.[7] Ventriculoperitoneal shunting is associated with a 70% incidence of complications that include status epilepticus, intraventricular hemorrhage, and subdural hematoma or hygroma.[20]

VI. Endovascular surgery

Endovascular treatment can use the transarterial, transvenous, transtorcular, or transvenous retrograde arterial approaches or some combination, but the most favorable route is transarterial since the risk of immediate or delayed hemorrhagic complication is significantly less with this approach.[6-8] When the transarterial route is not feasible, the transvenous approach is an alternative.[14,17] Transumbilical venous or arterial approaches are other possibilities.[4] A transumbilical venous approach enables both transarterial and transvenous embolization. Since the ductus venosus is usually closed at between 3 days and 1 week of life in neonates,[3] it is necessary to keep the umbilical vein patent at birth to use these approaches.

Transarterial embolization can be done following a diagnostic angiography using a microcatheter and a 4 French guiding catheter through a femoral sheath. Systemic heparinization is not necessary. Gentle navigation of the microcatheter to the target vessels is necessary because the wall of the feeding arteries is extremely thin. Transarterial embolization of the mural type of vein of Galen aneurysmal malformation can be done occasionally in one session, but it should be staged in most cases, especially in the choroidal type, since even partial occlusion of the arteriovenous shunt is often effective to improve clinical symptoms in neonates. Subsequent embolization to achieve a complete cure can be done later when the patient's general condition has improved.[8]

Transvenous embolization, either transfemoral or transtorcular, enables access to the dilated midline venous structure directly. Embolic material for this approach is usually coils. It is imperative to avoid migration or inappropriate placement of the coils. For this purpose, interlocking detachable coils provide excellent controlled delivery of the coils.[15] For the transtorcular approach in neonates, the bone is penetrated with a No. 18 needle and catheterization of the varix is performed without the need for a burr hole. However, this approach may cause perforation of the varix with a guiding catheter, resulting in fatal hemorrhage.[14]

Selection of the appropriate embolic material depends on the morphology of the lesion, the route of embolization, volume of shunted flow, accessibility to the vessel, and experience. We used interlocking detachable coils instead of glue. Glue might have migrated to the venous sinus and the pulmonary system due to the enormous arteriovenous shunt flows. Skillful experience is required to use glue for high-flow shunt without stenosis at the end of the ectatic vein.[7]

VII. Prognosis

Overall mortality in neonatal patients with vein of Galen aneurysmal malformation is 91% (64 of 70 cases) regardless of the treatment mode.[11] Most patients die within 1 week of diagnosis or treatment. This is the worst outcome among all age groups.[10,11] However, recent advances in modern endovascular technology have achieved a better outcome with long-term follow-up.[6-8,14] Secondary brain damage may occur due to abnormal brain circulation. Histological findings in neonatal patients are cerebral infarction *in utero*, periventricular leukomalacia, cortical laminar necrosis, subarachnoid hemorrhage, and hemorrhagic infarction.[18,23] Even after successful management, the functional prognosis is extremely poor, invariably with severe mental retardation.[18] Therefore, neonates with severe brain damage are not generally considered candidates for aggressive treatment.[7,8] Our patient had an intraventricular and subependymal hemorrhage, which was not thought to be due to birth trauma because the cesarean section was performed uneventfully. We decided to perform endovascular surgery on our patient in the belief that it would minimize global ischemic brain damage, thus improving functional prognosis and increasing the possibility of normal brain maturation.

Our patient died finally of pulmonary hemorrhage despite improvement of the congestive heart failure. The true cause of the pulmonary hemorrhage was unknown, but possible explanations include: persistent pulmonary hypertension that easily resulted in

pulmonary hemorrhage, or an adverse effect from the heparin that was used to keep the arterial and venous lines patent. Pulmonary hypertension was basically caused by immaturity of the lungs. Aggressive medical treatment for pulmonary hypertension and waiting for the lungs to mature are possible solutions. Strictly limiting the heparin dosage is also mandatory.

Whatever treatment modality is chosen, close cooperation between neurointerventionalists, neurosurgeons, neonatologists, anesthesiologists, and obstetricians is required to treat this challenging disease.[8,12]

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Address reprint requests to: M. Komiyama, M.D., Department of Neurosurgery, Osaka City General Hospital, 2-13-22 Miyakojima-hondohri, Miyakojima-ku, Osaka 534, Japan.