Endovascular Treatment of Dural Sinus Malformation With Arteriovenous Shunt in a Low Birth Weight Neonate
—Case Report—

Masaki KOMIYAMA, Yasuhiro MATSUSAKA, Tomoya ISHIGURO*, Shouhei KITANO*, and Hiroaki SAKAMOTO*

Abstract

A boy was born at 36 weeks gestation weighing 2,135 g, with a prenatal diagnosis of dural sinus malformation with arteriovenous shunts. Congestive heart failure and anuria at birth prompted emergency intervention. Transfemoral-transvenous coil embolization was performed on day 1, resulting in partial occlusion of the huge venous pouch with a total length of 2,355 cm of detachable coils. Transarterial glue embolization on days 7, 23, and 42 was required due to persistent heart failure. Transarterial embolization was performed by common carotid puncture because the transfemoral route could not be used due to the small size and compromised blood flow of the femoral artery. Transarterial embolization reduced the arteriovenous shunts markedly and resulted in clinical improvement. Early treatment of a high flow dural arteriovenous fistula in a low birth weight neonate can achieve an excellent result with an acceptable neurological outcome.

Key words: cerebral angiography, dural sinus malformation, embolization, heart failure, low birth weight, neonate

Introduction

Congenital dural arteriovenous fistulas in children are extremely rare. The presentation in childhood may involve cranial bruit, heart failure, dilated scalp veins, macrocephaly, hydrocephalus, delayed neurological development, seizure, and focal neurological deficits, which are distinct from the adult symptoms. Congenital dural arteriovenous fistulas can be divided into three subgroups: dural sinus malformation (DSM) with arteriovenous shunts (AVS), infantile-type dural AVS, and adult-type dural AVS. Pediatric dural AVS is characterized by a large, patent dural sinus with no venous lakes. Adult-type dural AVS is usually located in the cavernous sinus. DSM with AVS is observed in all age groups, but usually in neonates. DSM with AVS involves giant dural pouches and slow-flow mural AVS. Treatment of DSM with AVS in the neonatal period is challenging since the general condition is usually poor and/or vascular access is limited. We treated a low birth weight neonate harboring DSM with AVS using transfemoral venous and direct common carotid puncture endovascular treatment because of limited arterial access.

Case Report

A neonatal boy was the first child of healthy parents. Routine bi-weekly obstetric ultrasound examination identified an abnormal intracranial mass at 26 weeks gestation. The mother of the patient was referred to us at 36 weeks gestation for delivery and management of the child. Magnetic resonance imaging performed 1 day before delivery showed the lesion with a large flow void area, which was consistent with a diagnosis of DSM with AVS (Fig. 1).

Elective caesarian section under spinal anesthesia was performed at 36 weeks and 6 days gestation. No fetal distress was observed before delivery. Birth weight was 2,135 g and head circumference was 34.5 cm. Apgar scores were 5 and 7 at 1 and 5 minutes after birth, respectively. Endotracheal intubation was performed 1 minute after birth and mechanical ventilation was soon required. Chest radiography showed cardiomegaly with a cardio-
Fig. 1 T₂-weighted magnetic resonance image 1 day before delivery showing a huge mass lesion in the left parieto-occipital region. The lesion includes a flow void, which is consistent with a diagnosis of dural sinus malformation with arteriovenous shunts.

The thoracic ratio of 71% (Fig. 2A). The patient was neurologically normal. Bruits were heard over the head and in the cervical regions. Umbilical arterial catheterization failed, so umbilical venous catheterization was performed using a 4-French nutritional tube for possible transumbilical embolization.

Ten hours after birth, left retrograde radial angiography using 3 ml of contrast media was performed through a radial arterial route, which was also used for continuous arterial pressure monitoring, to plan embolization strategy. Angiography showed DSM with AVS. The huge dural pouch was located in the left parieto-occipital region. The meningeal branch of the left vertebral artery contributed to the DSM, but the presumed main feeding arteries from the left external carotid artery was barely visualized through fine collaterals (Fig. 2B, C). Even with the limited angiographical information, we were confident that the main feeding arteries originated from the left carotid arterial tree. Magnetic resonance angiography was not performed because of the high risk for a ventilated neonate with severe heart failure. Urgent reduction of shunt flow was necessary because of anuria and severe heart failure (heart rate over 220 beats/min). Transvenous coil embolization was scheduled on day 1 (24 hours after birth).

The transumbilical-transvenous approach failed because the catheter did not enter the inferior vena cava through the ductus venosus. Therefore, transfemoral-transvenous coil embolization was performed using detachable platinum coils (Detach-18 system; Cook, Bjaeverskov, Denmark). Only partial occlusion of the huge venous pouch was accomplished despite depositing a total coil length of 2,355 cm (Fig. 2D). Transvenous embolization resulted in slight clinical improvement and minimum urine output. Hemostasis at the femoral venipuncture site required about 90 minutes due to thrombocytopenia (platelet count 19,000/mm³), for which platelet transfusion was required.

Further reduction of the AVS was required to improve heart, respiratory, and hepatic failure. The arterial flow in the femoral artery was severely compromised and the artery was small, so the transfemoral-transarterial approach was contraindicated. Surgical exposure of the left common carotid artery for direct puncture and transarterial embolization were scheduled on day 7. Percutaneous puncture was avoided because of continued thrombocytopenia...
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Neurol Med Chir (Tokyo) 44, December, 2004

Platelet count 65,000/mm³ and the risk of inadvertent puncture of an arterialized jugular vein. Transfusion of platelets and red blood cells was intermittently required for thrombocytopenia and anemia. The distal common carotid artery was exposed in the neck, an 18-gauge plastic needle was inserted into the common carotid artery, and the tip was advanced to the proximal portion of the external carotid artery. Control angiography showed the huge dural pouch fed by large branches of left middle meningeal artery and occipital artery (Fig. 3). The three large branches of the middle meningeal artery and occipital artery were embolized with glue (30–40% mixture of n-butyl cyanoacrylate and lipiodol) through a microcatheter. Embolization resulted in marked reduction of the AVS and adequate urination due to increased renal flow. However, persistent congestive heart failure prompted the third embolization.

Angiography provided no information about the right carotid arterial system, but we thought that the right middle meningeal artery and occipital artery were contributing to the DSM. The right distal common carotid artery was exposed and direct puncture was performed as for the left side on day 23. The right middle meningeal artery and occipital artery were embolized with glue. Embolization resulted in only a small reduction of the AVS, but heart failure improved markedly. However, the patient could not be weaned from mechanical ventilation, and the fourth embolization was required.

The left distal common carotid artery was directly punctured using the previously opened left cervical skin wound on day 42. The cerebral feeding arteries were embolized using a flow-directed microcatheter. The three large feeding arteries of the anterior and middle cerebral arteries were embolized with glue (Fig. 4). His heart and respiratory failure further improved. Mechanical ventilation was discontinued and the patient was extubated on day 43.
Improved systemic condition allowed increased milk intake. Consequently, his body weight increased steadily from 2,168 g on day 45, to 2,492 g on day 61, and then 2,774 g on day 78. Magnetic resonance imaging of the brain on day 72 showed the moderately enlarged ventricular system and markedly shrunken dural pouch, which was partially thrombosed (Fig. 5). The patient was discharged from our hospital on day 99 when his body weight was 3,099 g. Cardiac echography at discharge showed normal cardiac function without volume overload. The patient underwent ventriculoperitoneal shunt for hydrocephalus at age 8 months and had minimal right hemiparesis and moderate developmental delay (developmental quotient 59) at the last follow-up examination at age 19 months.

Discussion

Neonatal cerebral vascular lesions causing high-output heart failure are usually vein of Galen aneurysmal malformations or congenital dural arteriovenous fistulas (mostly DSM with AVS), and less frequently pial arteriovenous malformation. DSM is rarer than vein of Galen aneurysmal malformation.14,17) No severe heart failure was observed in 19 neonates with DSM with AVS.17) However, DSM with AVS can cause severe heart failure immediately or soon after birth,6,10,11,14,19,26) as in our patient. Prolonged cerebral venous hypertension may cause brain damage, which will appear as dystrophic parenchymal calcification and white matter abnormalities. Since the brain parenchyma in neonates with DSM is usually normal,11) aggressive treatment is justified before brain damage occurs.

Embolization combined with medical treatment (diuretics, inotropic agents, and respiratory support) is now considered the treatment of choice for DSM with AVS because of its effectiveness and less invasiveness,11,17) although surgical treatment in conjunction with embolization has been successful in a few cases.6,21) Embolization can use the transarterial or transvenous including trans-torcular and trans-superior sagittal sinus (fontanelle) approaches, so embolization proceeds without knowledge of the angioarchitecture. Arterial access is usually required even for transvenous and direct access because control angiography is necessary to disclose the angioarchitecture.

Arterial access for embolization in children is usually limited to the femoral artery. A large AVS in the brain is associated with extremely decreased arterial flow in the descending aorta and prominently reversed aortic flow in the diastolic phase due to the steal phenomenon. Consequently, the flow to the femoral artery is extremely compromised even if the systolic pressure is normal. Therefore, the transfemoral approach in a low birth weight neonate (<2,500 g) poses a completely different situation from that in a normal birth weight neonate (>2,500 g). In a large series of patients with vein of Galen aneurysmal malformation, the smallest patient treated by the transfemoral approach weighed 2,800 g.16) Even in normal weight neonates, the femoral artery is small and insertion of a catheter or vascular sheath might cause severe ischemic and/or thromboembolic complications of the leg.2,5,22) We believe that insertion of a 4-French vascular sheath to the femoral artery might cause serious ischemic complications in low birth weight neonates.

Umbilical access, like femoral artery access, may allow both transarterial and transvenous embolization.2,14) Umbilical venous access allows both transvenous embolization and transcatheter (through the foramen ovale) transarterial embolization.14) However, the transumbilical approach is allowed only early in the neonatal period, and catheterization of the umbilical arteries and vein is not always feasible.12,24)

Another possibility is a direct access to the cervical carotid arteries, as used in our patient. Increased blood flow to the brain usually results in enlargement of the carotid arteries. Percutaneous puncture might be possible, but we chose direct puncture of the common carotid artery under direct vision after surgical exposure because we were afraid of inadvertent puncture of the arterialized jugular vein and resultant local hematoma. Furthermore, coagulation system disorder is occasionally observed in neonatal vascular lesions with AVS,7,11,15,25,26) as in our patient. Therefore, percutaneous puncture should be avoided.

Other possible approaches to the lesion include transfemoral-transvenous or transjugular-transvenous approaches, puncture of the fontanelle, and direct access via a small craniotomy above the lesion.3,7,18,26) Control angiography cannot be performed with these approaches, so embolization proceeds without knowledge of the angioarchitecture and hemodynamics. In this situation, real-time ultrasonography examination might be useful, but careful interpretation is necessary due to slow flow in the DSM.20)

Early endovascular treatment of DSM with a large AVS, even in a low birth weight neonate, can achieve an excellent result with an acceptable neurological outcome.
References


Address reprint requests to: M. Komiyama, M.D., Department of Neurosurgery, Osaka City General Hospital, 2–13–22 Miyakojima–Hondohri, Miyakojima–ku, Osaka 534–0021, Japan.

E-mail: komiyama@japan-mail.com