Transarterial Embolization of Congenital Dural Arteriovenous Shunt at the Torcular Herophili

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ABSTRACT

Congenital dural arteriovenous shunt (DAVS) is a rare disease entity. A neonate diagnosed to have cystic mass in posterior fossa during prenatal sonography presented with heart failure and hydrocephalus. Cerebral angiography revealed a DAVS at the torcular herophili. Successful occlusion of feeders was achieved by staged transarterial embolizations, with subsequent improvement of heart failure. Transarterial embolization could be a successful treatment method in pediatric patients with congenital DAVS, in case of limited numbered feeders. (Kor J Cerebrovascular Surgery 8:124-7, 2006)

KEY WORDS: Congenital intracranial arteriovenous malformations · Arteriovenous shunt · Torcular herophili · Therapeutic embolization

Introduction

Congenital dural arteriovenous shunt (DAVS) is an extremely rare disease entity.7 It may present with systemic signs and symptoms with higher mortality rate compared with that of adults,4,9 thus early treatment in the course of disease is essential for the DAVS in pediatric patients.

The treatment strategies of DAVS are still controversial, but classic procedures such as endovascular embolization and/or surgical resection may be an option. Here, we describe a male infant with DAVS involving the torcular herophili who was successfully treated with transarterial embolization.

Case Report

A male neonate, who had been diagnosed to have cystic mass in posterior fossa and right heart enlargement during prenatal sonography at the gestational period of 33 weeks, was delivered with a birth weight of 3400 g at the gestational period of 35 weeks by cesarean section. Apgar scores were 8 and 9 at 1 and 5 minutes after birth, respectively, but he suffered from hypoxemia and tachypnea since birth. Chest radiography showed cardiomegaly with pulmonary congestion and he manifested high-output congestive heart failure on echocardiography. Transcranial Doppler sonography (Fig. 1) showed a large sonolucent mass that disclosed turbulent bidirectional flow within it, and magnetic resonance (MR) imaging of brain demonstrated aneurysmal dilatation of the torcular herophili with severe hydrocephalus due to mechanical compression of the enlarged torcular herophili.

Cerebral angiography was performed on the 5th day after birth, showing multiple arteriovenous fistulae draining directly into the massively enlarged torcular herophili. The left middle meningeal artery was the dominant arterial feeder (Fig. 2A), and other small supplies from meningeal branches of bilateral ascending pharyngeal arteries and occipital arteries were observed. Intracranial blood flow was relatively decreased due to the large amount of shunt and
venous outflow restriction caused by the hypoplastic venous structures and sinus system (Fig. 2B). Occlusion of the dominant feeder was attempted via transarterial approach promptly on the same day. Endovascular embolization with multiple detachable coils (Guglielmi detachable coils, Boston Scientific/Target Therapeutics) resulted in partial occlusion of main fistula, and additional embolization was done with 50% mixture of iodized oil (Lipiodol; Andre Guerbet, Aulnay-sous-Bois, France) and glue (N-butyl cyanoacrylate, Histoacryl; B. Braun, Melsungen, Germany). The initial embolization resulted in complete occlusion of the dominant fistula (Fig. 2C) with improved intracranial arterial blood supply. Chest radiograph showed decreased heart size with improvement of hypoxemia and tachypnea.

An attempt of second endovascular treatment was performed 3 months after birth for the residual shunt flow and persistent hydrocephalus. Residual arterial supply via the occipital artery was occluded by detachable coils, glue and 150-250 microns polyvinyl alcohol (PVA) particles (Contour; Boston Scientific, Mississauga, ON, Canada) (Fig. 3) resulting in near complete occlusion of the residual fistulae.

The patient underwent follow-up angiography at the age of 9 months. Cerebral angiogram showed no residual shunt into the torcular herophili and venous outflow obstruction was improved (Fig. 4). Obstructive hydrocephalus was markedly improved, although it was persistent. He presented no more symptoms associated with the heart failure, and showed the normal development stage of two to three months-old.

**Discussion**

Dural arteriovenous shunts (DAVS) are rare disease that
account for less than 10% of all arteriovenous malformations of central nervous system and DAVS in neonates or infants are generally assumed to be congenital. Congenital DAVS are extremely rare and few cases are reported in the literature since the first case was reported by Epstein et al. in 1962. \cite{1,2,4,7,8}

Lasjaunias et al.\cite{5} divided DAVS in the pediatric ages into 3 subgroups hypothetically. These are dural sinus malformation with arteriovenous shunt (neonatal type), infantile (or juvenile type) DAVS, and adult-type (or adolescent type) DAVS. The dural sinus malformations with arteriovenous shunts (neonatal type DAVS) are found in neonates, showing giant dural lake and slow-flow communication with other sinuses and cerebral veins. Most of these lesions involve posterior dural sinuses or jugular bulb. Angioarchitecture of infantile type DAVS is large and patent dural sinus without venous lakes. High flow rate of shunt and multifocality are main characteristics of infantile DAVS. Adult-type DAVS are usually located in cavernous sinus or sigmoid sinus region and found in older children. This case involved posterior dural sinus with giant dural lake formation, so it can be included in the neonatal type DAVS.

DAVS in neonates or infants manifest systemic signs and symptoms, which differentiate congenital DAVS from adult one, such as macrocephaly with hydrocephalus, seizure, delayed neurological development and cardiac failure. And mortality rate of DAVS is higher in neonate (67%) than in other age groups (31~38%),\cite{4,8} thus early treatment is mandatory in this group. They show relatively poor prognosis, especially when venous infarct with hemorrhage occurs.\cite{5} This case does not show venous infarct or hemorrhage until last follow-up, but torcular herophili involvement can be another poor prognostic factor.\cite{5}

The therapeutic strategy for DAVS is endovascular embolization or surgical resection of the involved sinus. In neonates and infants, surgical elimination of dural fistulas is more hazardous and difficult than that in adults. In the review of Morita et al.,\cite{8} such lesions carry a poor prognosis with a reported mortality of 38%, and with a historical anatomic cure of only 9%. Therefore, endovascular

![Fig. 3. Left common carotid angiogram with lateral view during the second embolization on the 3 months after birth. A: Arterial phase shows shunt via left occipital artery arrow and increased intracranial blood flow compared with Fig. 2A. B: Left occipital artery was occluded by detachable coils, glue and polyvinyl alcohol particles.](image)

![Fig. 4. Left common carotid angiogram with lateral view at the age of 9 months during follow-up. A: Arterial phase shows no residual shunt into the torcular herophili. B: On venous phase, superior and inferior sagittal sinuses, inferior petrosal sinus, superficial cortical and deep cerebral veins are visualized, which were not seen in Fig. 2B.](image)
embolization combined with medical treatment (diuretics, inotropic agents, and respiratory support) is now considered the treatment of choice for DAVS because of its effectiveness and less invasiveness. Endovascular treatments for DAVS are usually performed via transarterial or transvenous approach. As a rare case, Komiyama et al. reported transumbilical embolization of congenital DAVS at the torcular herophili in a neonate and Liu et al. reported transtorcular embolization through a needle-sized craniotomy.

In our case, alternative venous outflow was not developed apparently, and embolization via transvenous route was not allowed. Feeders were relatively limited in number and repeated transarterial embolizations were performed, which might be the cause of successful embolization in this patient.

Conclusion

We reported the case of a neonate with a rare congenital DAVS at the torcular herophili. This patient was treated by staged transarterial embolization, and transarterial embolization can be a successful treatment method in case of limited numbered feeders.

REFERENCES