Case Report

Dural sinus malformation with arteriovenous fistulae in a newborn: Positive outcome following endovascular management


1. Introduction

Dural sinus malformation (DSM) is a rare intracranial vascular malformation that presents in newborns and is characterized by a dilated dural sinus and associated with dural arteriovenous fistula (DAVF). The typical clinical manifestations are similar to intracranial high-flow arteriovenous fistulas that often present with hyperdynamic heart failure and macrocrania. The clinical outcome of DSM is largely dependent on its location and severity. Patients usually have favorable outcomes when the DSM is localized in the unilateral dural sinus with small and/or slow DAVF. On the hand, poor neurological prognosis occurs if the DSM involves the large/central dural sinus, such as in torcular herophili and/or superior sagittal sinus (SSS). Here, we present an unusual case of DSM that involved the SSS, torcular herophili, and left transverse sinus that was successfully treated by endovascular management.

2. Case report

A full-term baby girl was born to a 40-year-old nulliparous mother who was being medically treated for hyperuricemia. The mother maintained alcoholic drinking habits during pregnancy. The mother had stopped taking her medication during pregnancy and had not received regular obstetric ultrasounds. No fetal distress was observed before or after delivery. The infant’s birth weight was 2623 g, with mild enlargement of the head (head circumference: 35 cm) noted at birth. The baby was discharged 10 days later under stable conditions. However, several seizures occurred on Day 14 after delivery, with tremors in all four limbs occurring for several seconds approximately every 2 hours. During hospitalization, her seizures were well controlled by anti-epileptic drugs. Chest radiography indicated cardiomegaly, with a cardiothoracic ratio of about 75%. Due to mild enlargement...
of the head and the history of seizures, brain computed tomography (CT) was performed, which showed a relatively large, high-density lesion in the posterior fossa along the SSS (Fig. 1). Vascular anomalies, including arteriovenous malformations, were also indicated. Magnetic resonance imaging (MRI) demonstrated a large vascular pouch located at the torcular herophili and posterior SSS, consistent with the diagnosis of DSM.

Transfemoral cerebral angiography was performed under general anesthesia in order to further delineate the angioarchitecture of the DSM. Angiography demonstrated a large, dilated vascular pouch in the posterior SSS and torcular herophili and left transverse sinuses associated with DAVFs; the main feeders of the DAVFs were the bilateral middle meningeal arteries (MMAs), bilateral occipital arteries, and the right posterior cerebral artery (Fig. 2). Transarterial embolization was performed after cerebral angiography. A coaxial microcatheter (SL-10; Boston Scientific Inc, Fremont, CA, USA) with a 4-F guiding catheter was navigated through two branches of the right MMA near the fistula. Then, a total of 1.2 cc of liquid adhesives, composed of a 50/50 mixture of N-butyl-2-cyanoacrylate (NBCA; Nycomed, Igenor, Paris, France) and Lipiodol Ultra-Fluide (Laboratoire Guerbet, France), was injected into the two fistulae through the right MMA. Because of limitations in the amount of available contrast media in this low body weight infant, further embolization was not performed. The postembolization clinical course was uneventful. Improved systemic conditions without any seizure episodes and increased milk intake (from 40 mL every 3 hours to 60 mL) were noted in the following 2 months. The infant’s body weight also increased steadily from 2912 g on Day 30 to 4496 g on Day 99. A second embolization session was initiated on Day 100 in order to further occlude the fistula flow. Pre-embolization cerebral angiography showed a marked reduction in the size of the vascular pouch with normalization of the left transverse sinus. The residual DAVFs were fed by the bilateral occipital arteries and the right posterior cerebral artery (Fig. 3). Embolization with occlusion of the arteriovenous fistula, which was fed by the right posterior cerebral artery, was achieved following an injection of 0.5 cc of the NBCA mixture (Fig. 4). A postembolization CT scan showed a reduction in the size of the vascular pouch (Fig. 5) and regression of the white matter abnormalities. The patient was discharged on Day 111 of hospitalization with a body weight of 4776 g and a normal daily food intake. Long-term follow-up imaging examinations were not performed. However, on a clinical follow-up examination performed at the age of 2 years, she presented with normal neurological development without delays in any neurological milestones.

3. Discussion

The most common pediatric intracranial AV shunt is an aneurismal malformation of the vein of Galen (VGAM) (41%). DSM is uncommon and accounts for less than 2% of vascular malformations. Clinically, DSM mimics other intracranial high-flow fistulae that present in infants because it shares many similar clinical symptoms with early cardiac failure and can result from high cardiac output and/or macrocrania, particularly in infantile dural arteriovenous fistula (IDAVF). However, DSM presents with a dilated or giant pouch in the dural sinus that is usually associated with a low
and/or relatively slow flow through the dural AV shunt. On the other hand, in IDAVF, the sinus usually presents with a normal or slightly enlarged dural sinus with high-velocity flow through the AV shunt. In addition to the morphology of the dural sinus, DSM usually presents with acute symptoms such as convulsions, venous reflux, infarction, or hemorrhage, while IDAVF manifests with marocrania, mental retardation, or progressive neurological deficits depending on the location of the occluded secondary sinus.3

The exact mechanism that results in the formation of DSM remains unclear. Okuda et al reported that underdevelopment of the jugular sinus, which serves as a blood reservoir, results in the ballooning of the transverse sinuses and posterior superior sagittal sinus to occur at 4–7 months of intrauterine age. The transverse sinus gradually remodels during the postnatal stage as the jugular bulb forms starting at 2 years of age.4 DSM had been proposed to occur due to the ongoing ballooning of the transverse sinus. However, this has not been reported in normal neonates and cannot explain the occurrence of AV shunts in the sinus wall. It is now believed to be associated with the uncontrolled development of the posterior sinuses, including the transverse, sigmoid, and confluent sinuses.2

The natural course of DSM depends on the location and extent of the dural sinus and the fistula flow, as well as the timing of any endovascular treatments that are prescribed. In previous studies,2,3 poor prognosis was reported when DSMs involved immature, large, or central dural sinuses with turbulent and slow flow and impairment of the deep and superficial veins. In these circumstances, spontaneous thrombosis of the dural sinus, venous hypertension, and irreversible brain damage, including venous infarction and intracranial hemorrhage (ICH), may occur. Lasjuaia et al reported that 64% of patients die after receiving conservative treatment.1 On
the other hand, when the DSM is localized in a unilateral dural sinus (e.g., lateral sinus or jugular bulb) without affecting the torcular herophili or SSS, a better clinical prognoses have been observed because the brain could be drained via the contralateral sinus and other anastomosis even if the involved sinus is thrombosed or occluded.\textsuperscript{4,5}

The treatment strategies for DSMs involving large or central dural sinuses include gradual closure of the fistula flow in order to relieve intracranial venous hypertension and prevent cardiac failure. Sudden and rapid occlusion of all the flow through the fistula in a single session may carry the risk of brain hyperperfusion and/or dural sinus thrombosis, which can lead to catastrophic complications such as venous ischemia, infarction, or hemorrhage. Therefore, performing endovascular embolization in multiple stages is critical to achieving a balance between reducing the impact of the flow through the fistula to the brain and heart while still maintaining the patency of the dural sinus and allowing the abnormal dural sinus time to undergo normal remodeling. In addition, medical treatment with heparin, diuretics, and/or inotropic agents help to prevent dural sinus thrombosis and hyperdynamic cardiac failure. Surgical treatment alone without previous embolization carries the risk of significant intra-operative blood loss with high rates of mortality and morbidity, which is of particular concern when treating infants and neonates.\textsuperscript{6} Positive clinical outcomes following surgical treatment in combination with embolization has been reported in neonates.\textsuperscript{7,8} Endovascular embolization in combination with symptomatic treatment remains the primary therapeutic method. Radiation therapy has been used to treat adults with localized, slow flowing dural lesions, but there are no such reports about using similar treatment on neonates, largely because of the risks of adverse effects.

In this particular case, the markedly dilated left transverse sinus, torcular herophili, and posterior SSS were depicted and fed by the bilateral middle meningeal arteries, bilateral occipital arteries, and the right posterior cerebral artery. There was no evidence of venous strain, such as jugular vein stenosis or pial reflux. Aggressive management by direct occlusion of the SSS or torcular herophili is hazardous and may lead to venous hypertension, infarction, or hemorrhage. Instead, we only embolized the feeders to the right MMA in the first session of treatment because of the limited amount of contrast medium required for this low-bodyweight infant. However, spontaneous occlusion of the majority of the DAVFs fed by the middle meningeal and occipital arteries was demonstrated at a 2-month follow-up examination; furthermore, gradual regression and normalization of the involved dural sinus were found. In addition, white matter abnormalities in the parieto-occipital region that were discovered on pretreatment MRI were regressive.

Previous studies have proposed that brain damage results in a poor clinical prognosis.\textsuperscript{1} In this particularly case, however, some brain damage to the parieto-occipital region occurred with minimal neurological deficits. The patient had a favorable clinical outcome and good recovery in terms of the lack of neurological deficits that were noted during the follow-up period. This is different than previous reports on DSMs involving SSS and torcular herophili,\textsuperscript{3,9} largely because the patient had patent intracranial venous drains and relatively slow fistula flows. Another important key factor that prevented severe brain damage was the early application of embolization in order to control the fistula flow before the occurrence of irreversible brain damage and to facilitate the subsequent remodeling of the venous system.

DSM is a rare, congenital, intracranial, dural, arteriovenous malformation that may be associated with a poor clinical prognosis, particularly when DSM involves the central, large dural sinus and sinus confluence. Early diagnosis with early, staged endovascular treatment to control fistula flow is crucial for the preservation of enough venous drainage of the brain and to diminish the impact of the fistula flow to the brain and heart.

\section*{References}


