Endovascular treatment for pediatric intracranial arteriovenous shunts

Yasunari Niimi, MD, PhD
Department of Neuroendovascular Therapy, St. Luke's International Hospital
Tokyo, Japan

Abstract

Intracranial arteriovenous shunts (ICAVSs) in young children are characterized by high frequency of high-flow fistulas. In association with physiological condition of the developing brain and the heart, intracranial high flow fistulas in young children present with unique symptoms at specific ages. It is also noteworthy that ICAVSs in this age group are often associated with genetic or congenital disorders. In neonate, vein of Galen aneurysmal malformation (VGAM) and dural sinus malformation (DSM) with arteriovenous shunts (AVSs) tend to present with high output cardiac failure. In infancy, VGAM, pial arteriovenous fistula (AVF), and infantile dural AVF (DAVF) tend to present with hydrodynamic disorders such as macrocephaly, ventriculomegaly, prominent facial veins, and developmental delay. Pial AVF, nidus type pial arteriovenous malformation (AVM), and infantile DAVF can present with focal neurological deficits, seizure, or hemorrhage at an older age. Endovascular treatment is currently the first choice of treatment for most of pediatric ICAVSs. Treatment goal should be set up based on the unique physiological condition of the children. For neonates, treatment goal is usually to control high output cardiac failure. For infants, hydrodynamic disorder is the main target to let the child develop normally. After 1 year old, anatomical cure of ICAVS can be aimed if possible. Staged multiple endovascular treatments may be necessary for complicated cases. I will review characteristic features of pediatric ICAVSs and outline their endovascular treatment with representative cases.

Classification

Classification of ICAVSs by their location in the space covering the brain is helpful for understanding the pathophysiology of the diseases (Table 1). Pial AVMs and AVFs exist in the sub-pial space, VGAMs exist in the subarachnoid space, and dural AVFs exist within the dural layer.

Presenting age and Symptoms

One of the major characteristics of pediatric ICAVSs is that each disease presents at a specific age with a specific symptom. This is related to the fact that most of ICAVSs in the early childhood are high flow fistulas. Existence of high flow shunting in a certain layer of the brain

coverage creates specific pathophysiology, associated with specific physiological condition of the developing brain and heart at a certain age.

Fetus

ICAVSs diagnosed in the fetus are mostly VGAMs or DSMs, and rarely pial AVFs. Not all the diseases diagnosed in the uterus should be treated during the neonatal period[1]. Some are out of indication for treatment because of already existing severe brain or organ damage with poor prognosis, and the others do not need to treat until the child grows older or becomes symptomatic.

Neonate

ICAVSs presenting in the neonatal period are also mainly VGAMs or DSMs, and most of them present with severe congestive heart failure (CHF). Pial AVF rarely presents in neonate, mostly with CHF but also with focal neurological symptoms such as seizure or hemorrhage. Neonates tend to develop severe CHF due to high flow AVF because they experience significant hemodynamic modification at birth, such as loss of placenta and increase in pulmonary circulation due to initiation of pulmonary ventilation, in addition to the volume overload by the high flow shunting.

For the neonatal high output cardiac failure, the goal of treatment is to control the cardiac failure to have a baby tolerate oral feeding and allow weight gain. If the baby can be discharged home by medical treatment, endovascular treatment is scheduled at approximately 5 months of age. If medical treatment is not sufficient to control cardiac failure, the patient needs emergency embolization in the neonatal period. The cardiac status of the patient usually significantly improves if the flow through the ICAVS can be decreased by approximately 50% by embolization[2].

Infancy

In infancy, cases of pial AVF and infantile type DAVF increase in addition to VGAM and DSM. Characteristic presentation at this age is hydrodynamic disorder, manifested as macrocephaly, ventriculomegaly, prominent facial veins, and developmental delay. Hydrodynamic disorder is a state of the brain with disturbed cerebrospinal fluid (CSF) absorption due to venous hypertension caused by ICAVS. It can occur in fetuses, neonates, infants, and young children but usually manifest in infants and young children.

With existence of high flow ICAVS, the sigmoid sinus and the jugular bulb tend to be occluded as a part of natural history, causing cortical reflux of venous drainage of the ICAVS

and aggravation of venous hypertension of the brain, resulting in seizures, hemorrhages, and neurological deficits. Chronic venous ischemia of the brain induces bilateral subcortical white matter calcification and brain atrophy, which can be manifested as psychomotor developmental delay. If the alternative pathway of the venous drainage of the brain from the cortical veins to the cavernous sinus is established by the time of sinus occlusion, the symptoms of venous hypertension is relatively mild. Endovascular treatment in time can prevent or even reopen occlusion of the jugular bulb[3].

Ventricular shunting reverses normal pressure gradient from the ventricle to the superior sagittal sinus. This causes brain edema, further enlargement of the dilated draining vein, calcification of the white matter starting opposite to the side of shunt placement, and subependymal atrophy. If venous hypertension is reduced early enough by endovascular embolization, hydrocephalus can be prevented or treated without ventricular shunt placement. Subacute progression of hydrodynamic disorder causes melting brain syndrome. In this situation, cerebral blood flow is decreased due to venous hypertension, and brain parenchyma is acutely and progressively destroyed mainly in the white matter. The ventricular system is enlarged without increased intracranial pressure. Melting brain syndrome can occur in all kinds of ICAVSs in fetuses, neonates, and infants, but not in adults.

Treatment goal for this age is to restore the normal hydro-venous equilibrium to permit normal development of the patient, avoiding ventricular shunt surgery.

Older children

After 2 years of age, new presentation of VGAM and DSM becomes rare, and pial AVM and AVF and infantile DAVF are seen more frequently. VGAM is rarely discovered in an older child with headaches, seizure, intracranial hemorrhage, or incidentally. Pial AVMs and AVFs tend to present with focal neurological manifestation such as hemorrhage or seizures, because they are in the subpial space as opposed to VGAM or DAVF which are extra-pial in location and tend to present with systemic symptoms (CHF) or symptoms of the entire brain (hydrodynamic disorder).

Endovascular treatment

Endovascular embolization is the first choice of treatment for most of the ICAVSs. Transfemoral transarterial embolization is our first and predominant choice of treatment. A trans-umbilical artery approach is preferable for newborn patients because of small size of the femoral artery. Transarterial embolization is performed using a flow guided microcatheter and N-butyl-cyanoacrylate (NBCA) as an embolic agent. High concentration NBCA mixture is injected under systemic hypotension to avoid migration of the mixture to the lung [4]. Coils can be used to close an aneurysm or venous side of the fistula. Onyx (ev3 Neurovascular, Irving CA)

is beneficial for DAVFs and certain BAVMs but not for high flow fistulas[5]. For high flow pediatric ICAVSs, DAVFs can develop naturally or following endovascular embolization. These DAVFs can be at the site of original ICAVS or remote from it[6].

Brief disease descriptions

Vein of Galen Aneurysmal Malformation (VGAM)

VGAM is a rare AVF of pediatric population and consists of less than 1% of the intracranial vascular malformation. They are located in the subarachnoid space in the velum interpositum cistern and quadrigeminal cistern. They are supplied by the choroidal arteries and drained by the embryonic draining vein of the choroid plexus, the median vein of Prosencephalon (Markowski). Additional supply can be seen from quadrigeminal arteries, thalamoperforators, and dural feeders. This embryonic vein exists between 6 and 11 weeks of gestation, and the posterior portion of this vein becomes vein of Galen as the internal cerebral vein develops to drain the deep brain structures. VGAM develops in relation to the arteriovenous system of the choroid plexus, which develops earlier than the arteriovenous system of the brain[7]. Therefore, the draining vein of the VGAM is, in principle, not connected with the cerebral veins. Some pial AVMs or AVFs draining to the vein of Galen can cause dilatation of this vein, which is the matured vein of Galen and has connection with cerebral veins including internal cerebral veins. These ICAVSs are called Vein of Galen aneurysmal dilatation (VGAD) and tend to present with hemorrhage, focal neurological deficits, or seizures at an older age than VGAM [8]. However, they can also present with CHF in neonate or with hydrodynamic disorder in infancy, whose radiological and clinical differentiation from the VGAM is sometimes difficult [1]. There are several case reports of VGAM which had co-existing internal cerebral vein draining to the dilated vein of the malformation.

Lasjaunias subclassified VGAM to the choroidal type (Figure 1) and the mural type (Figure 2), but there are cases of combination of both types[9]. The choroidal type is more primitive and has multiple complex fistulas along the elongated median prosencephalic vein through the arterial network. They tend to present with severe CHF during the neonate and more difficult to treat than the mural type. The mural type has one or few direct fistulas to the round shape dilated median prosencephalic vein, and tends to present with hydrodynamic disorder during infancy. I added an additional type, the quadrigeminal type, which tends to present at an older age than the mural type. Including this type, I am proposing a new classification of VGAM which should be called “vein of Galen AVF” regardless of existence of the internal cerebral vein (Figure 3, 4).

Brain AVM/ AVF

Brain AVM is subclassified to narrow sense pial AVM with a nidus and pial AVF

consisting of only arteriovenous fistulas without a nidus. Brain AVM is the most common cause of pediatric cerebral hemorrhage and 20% of them are diagnosed at younger than 20 years old. Eighty percent of pediatric brain AVMs present with hemorrhage with a higher mortality rate than adults. Characteristic features of pediatric brain AVM include 1) high frequency in the deep location, 2) high frequency of pial AVFs, 3) Common to see venous changes such as venous ectasia and thrombosis but rare to see arterial changes such as flow related aneurysms [8]. Pial AVF can be single fistula or multiple fistulas, and occur anywhere in the brain but more often above the tentorium. They can present with high output cardiac failure in the neonate or hydrodynamic disorder, seizure, or hemorrhage during infancy or older childhood[9]. Pediatric pial AVFs have high association with HHT and CM-AVM syndrome (Figure 5).

Dural AVF

DAVSs are subclassified into DSM, the infantile type, and the adult type.

DSM (Figure 6) is caused by mal-development and dilatation of the dural sinus and frequently involves the torcula and the transverse sinus. Mal-development of the dural sinus is the primary event in this disease and DSM without AVF can also occur. The dilated abnormal dural sinus tends to develop thrombosis inside. Depending on the location, flow through the AVF, and degree of the thrombosis, DSM can present with various symptoms such as CHF, hydrodynamic disorder, seizure, and hemorrhage mainly during neonate and infancy. This disease generally has a poor prognosis but those with preserved venous drainage of the brain due to the patent torcula or established alternative collateral pathways have a relatively good prognosis after endovascular treatment.

Infantile type is high flow fistulas which are often multiple in location and sometimes induce secondary pial AVFs. It is often difficult to treat because of extensive high flow AVFs and tendency to develop secondary fistulas. They tend to present with papilledema, seizures, and developmental delay during infancy and early childhood. Delayed thrombosis of the dural sinuses aggravates venous hypertension and then the hydrodynamic disorder as often seen in VGAM.

Adult type is an acquired lesion induced by triggering factors such as sinus thrombosis and trauma as seen in adult DAVFs. It is often seen in the cavernous sinus and has a relatively good prognosis[10].

References

1 Berenstein A, Niimi Y. Vein of Galen aneurysmal malformation. In: Winn R (ed.). *Youmans Neurological Surgery*. Vol. 2. Elsevier Saunders, Philadelphia, 2011; 2150-65.

2 Garcia-Monaco R, De Victor D, Mann C, Hannedouche A, Terbrugge K, Lasjaunias P.

Congestive cardiac manifestations from cerebrocranial arteriovenous shunts. Endovascular management in 30 children. *Childs Nerv Syst*. 1991; **7**: 48-52.

3 Berenstein A, Toma N, Niimi Y, Paramasivam S. Occlusion of posterior fossa dural sinuses in vein of Galen malformation. 2016; 37: 1092-1098

4 Berenstein A, Niimi Y, Song J, Lasjaunias P. Vein of Galen Aneurysmal Malformation. In: Albright A, Pollack I, Adelson P (eds.). *Principles and practice of pediatric neurosurgery*. 2 edn. Thieme, New York, 2008; 1014-28.

5 Niimi Y. Endovascular treatment of pediatric arteriovenous shunt. *Pediatrics International.* 2017; 59: 247-257

6 Paramasivam S, Niimi Y, Meila D, Berenstein A. Dural arteriovenous shunt development in patients with vein of galen malformation. *Interv Neuroradiol*. 2014; **20**: 781-90.

7 Raybaud CA, Strother CM, Hald JK. Aneurysms of the vein of Galen: embryonic considerations and anatomical features relating to the pathogenesis of the malformation. *Neuroradiology*. 1989; **31**: 109-28.

8 Lasjaunias P, Ter Brugge K, Berenstein A. Vein of Galen aneurysmal malformation. *Surgical Neuroangiography 3 Clinical and interventional aspects in children*. Vol. 3. 2 edn. Springer, Berlin Heidelberg, 2006; 105-226.

9 Paramasivam S, Toma N, Niimi Y, Berenstein A. Development, clinical presentation and endovascular management of congenital intracranial pial arteriovenous fistulas. *J Neurointerv Surg*. 2013; **5**: 184-90.

10 Lasjaunias P, TerBrugge KG, Berenstein A. Dural Arteriovenous Shunts. *Surgical Neuroangiography*. Vol. 3. Springer-Verlag, Berlin Heidelberg, 2006; 389-454.

Table 1: Classification of ICAVS (from ref 5)

Layer	Disease	
subpial	Pial AVM (nidus)	
	Pial AVF	
subarachnoid	VGAM	
dura	Dural AVF	DSM
		Infantile
		Adult
Extra-dural	AVM of bone, soft tissue, skin	

AVM: Arteriovenous Malformation, AVF: Arteriovenous Fistula, VGAM: Vein of Galen Aneurysmal Malformation, DSM: Dural Sinus Malformation

Figures

Figure 1

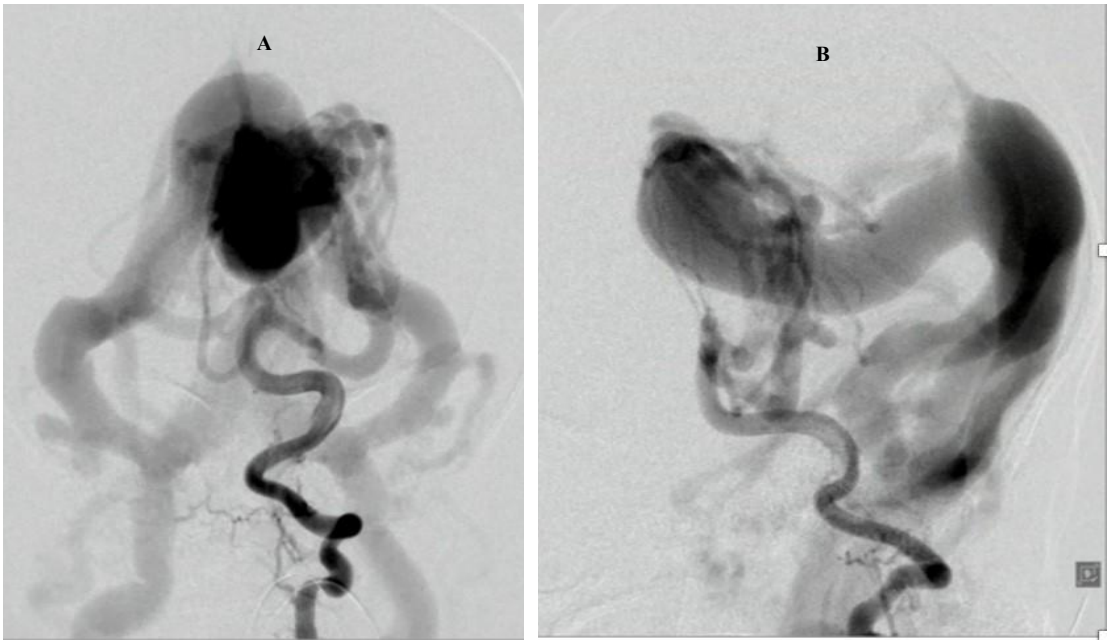

Figure 1: Choroidal type VGAM of a newborn girl. A, B: PA (A) and lateral (B) views of the left vertebral artery angiogram showing typical appearance of a choroidal type VGAM. Multiple feeders are supplying the VGAM draining to the embryonic falcine sinus. Bilateral occipital sinuses are patent.

Figure 2

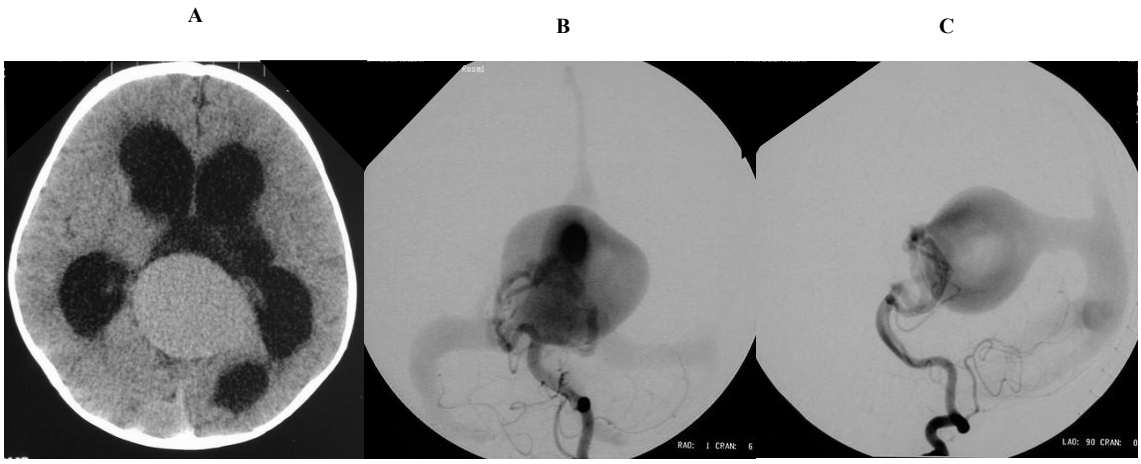

Figure 2: Mural type VGAM of a 6 month-old girl with macrocepahly. A: Non contrast CT showing enlarged vein of Galen and ventriculomegaly. B, C,: PA (B) and lateral (C) views of the left vertebral angiogram showing mural type VGAM supplied bilateral posterior choroidal arteries and drained by the embryonic falcine sinus.

Figure 3

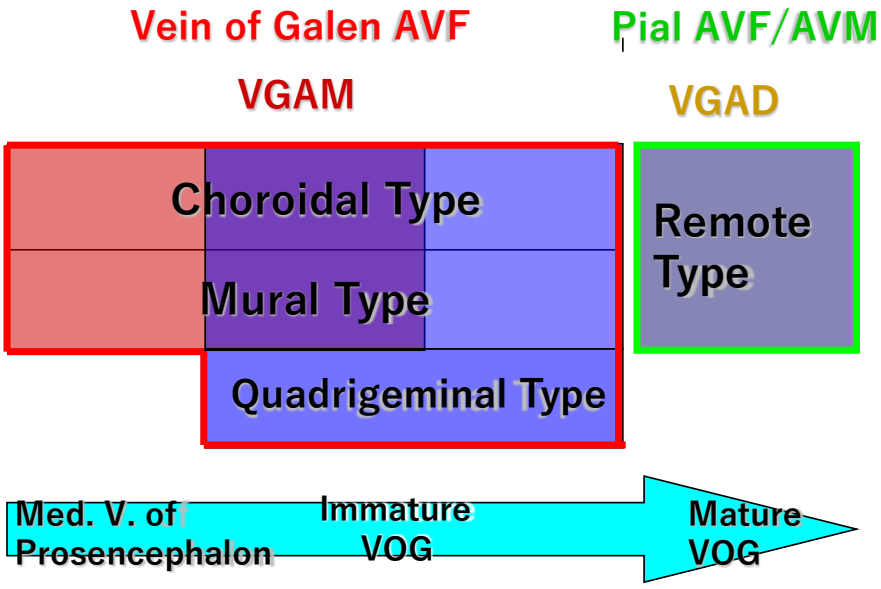

Figure 3: Proposed classification of VGAM regardless of existence of the internal cerebral vein.

Figure 4

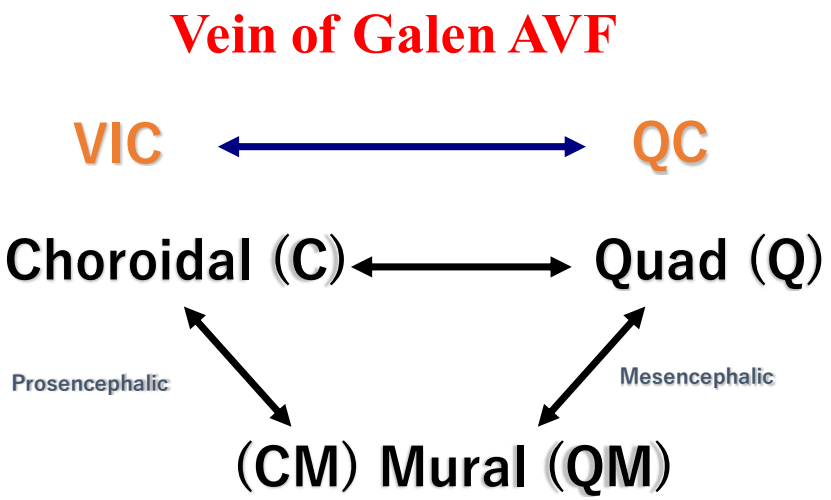

Figure 4: Locations and types of vein of Galen AVFs. VIC: verum interpositum cistern, QC: quadrigeminal cistern, C: choroidal, CM: choroidal mural, QM: quadrigeminal mural, Q: quadrigeminal

Figure 5

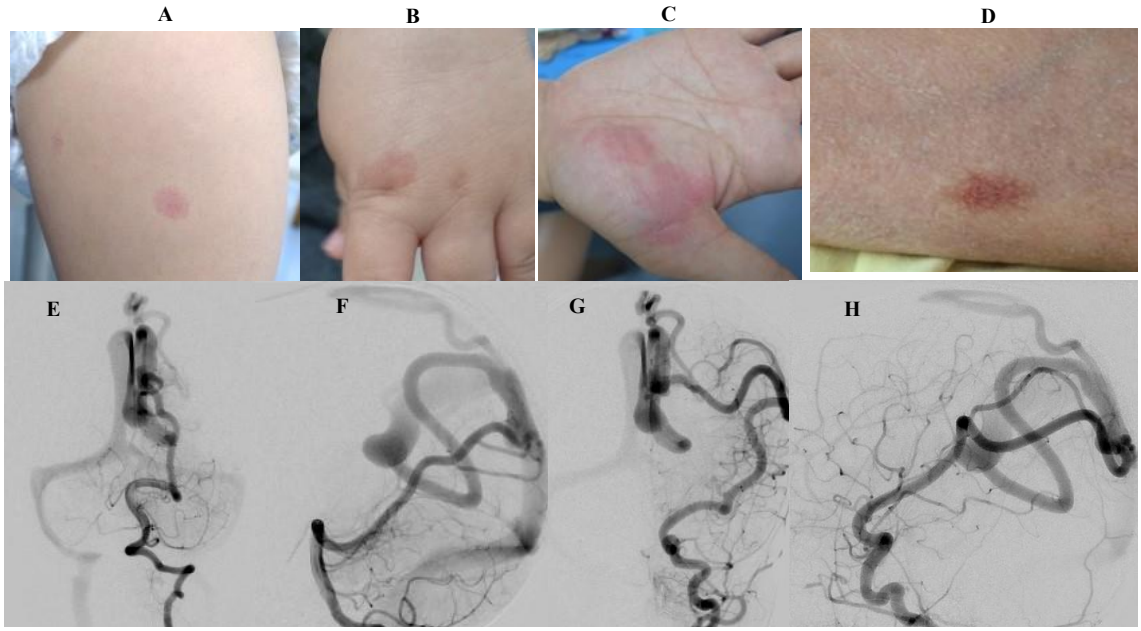

Figure 5: A-D: Clinical photographs of 11 months old boy (A), his younger sister (B), mother (D), maternal grandfather (D), showing typical capillary malformations of CM-AVM. They were genetically proven to be CM-AVM 1. E, F: PA (E) and lateral (F) views of the left vertebral angiogram. G, H: PA (G) and lateral (H) views of the left common carotid angiogram. There are multiple pial AVFs draining to the same cortical vein.

Figure 6

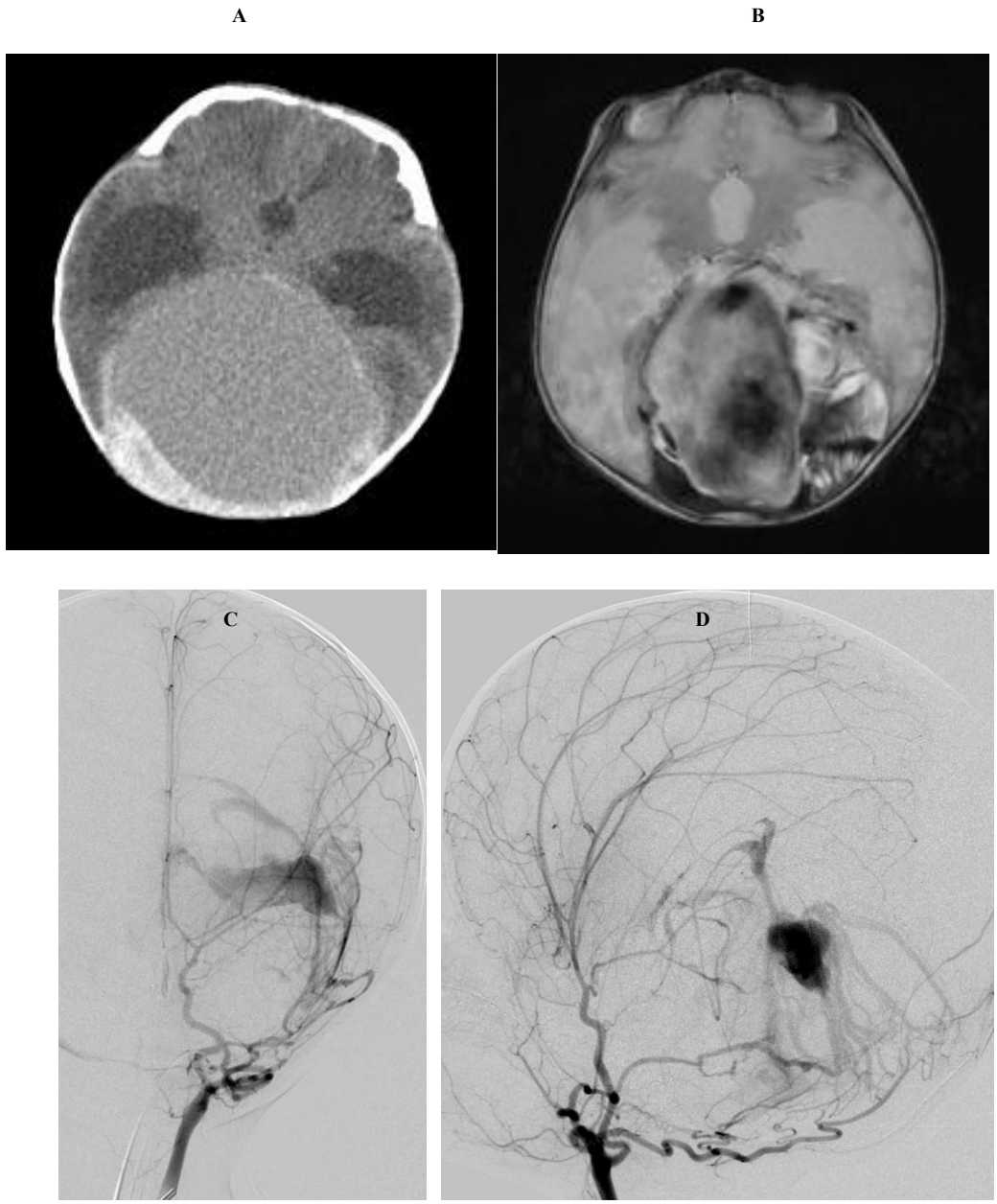

Figure 6: DSM of a newborn boy initially suspected a brain tumor by the intrauterine ultrasound and MRI.

A: CT at day 0 showing the large torcula and hydrocephalus.

B: Axial FLAIR image of MRI at day 5 showing progressive thrombosis of the large torcula, hydrocephalus, and bilateral cortical signal abnormality. C, D: PA (C) and lateral (D) views of the left common carotid angiogram obtained on day 20, showing dural AVSs to the large partially thrombosed torcula supplied by dural branches of the occipital, posterior auricular, middle meningeal, tentorial, and posterior cerebral arteries.