Infantile Dural Arteriovenous Shunt Draining into a Developmental Venous Anomaly

S. GEIBPRASERT, T. KRINGS*, V. PEREIRA, P. LASJAUNIAS

Diagnostic and Therapeutic Neuroradiology Service, Hôpital Bicêtre, Le Kremlin-Bicêtre, Paris, France

* Departments of Neuroradiology and Neurosurgery, University Hospital Aachen, Germany

Key words: pediatric dural arteriovenous shunt/fistula, developmental venous anomaly, cavernoma

Summary

A three-year-old girl with an incidentally discovered infantile type of dural arteriovenous shunts (DAVs) along the superior sagittal sinus during investigation of a minor head trauma is presented.

The DAVs drained into a developmental venous anomaly of the right cerebral hemisphere. In addition, there was a small cavernoma within the territory drained by the DVA. The patient underwent multiple transarterial embolizations to decrease the shunt flow and thus the constrained venous drainage of the DVA and right cerebral hemisphere. Pediatric dural arteriovenous shunts are a different entity from the adult type DAVs and should be managed according to the growth and development of the child. DVAs are extreme variations of the venous system with reduced flexibility to increased venous drainage. Regardless of the type of treatment employed, the DVA and its drainage pathway must be preserved.

Introduction

Dural arteriovenous shunts (DAVs) in children are a special and rare disease with a high overall mortality¹⁻⁶, reported to be 38% by Morita et Al.⁶ and up to 67% for neonatal cases. Three different types of the disease have been described, the most common found in the

literature being the infantile type^{3-5,7}. Associations with developmental venous anomalies (DVAs) are extremely rare and according to the time scheme for development of venous diseases and variations along the arterio-venolymphatic tree^{1,8}, are more likely related to the dural sinus malformations rather than the infantile type of dural arteriovenous shunts. Only four cases have been reported in literature^{5,8-10} of which there were only two in the pediatric population^{5,8} and only one case being of the infantile type.

Case Report

A three-year-old girl was referred to our hospital with incidentally discovered infantile type DAVs along the superior sagittal sinus during the investigation for minor head trauma after falling off a bicycle. On admission she was neurologically intact with a head circumferential of 53cm (+ 2SD, >98%). A persistent cranial bruit in the midline was noted.

The patient had an older sister with multiple anomalies of the limbs with cortical malformation. The mother's history was remarkable for two prior pregnancy terminations due to one chromosomal abnormality and one anencephaly, but in neither case was definite genetic diagnosis obtained. There was also a notable family history of interfamilial marriage of the previous generations.

MRI showed a large tubular structure (inferior to the right corpus callosum, connecting with the vein of Galen) along the course of the right internal cerebral vein, draining multiple dilated transmedullary veins from the right frontoparietal lobe, compatible with a DVA. Prominent cortical veins on both cerebral hemispheres were also noted. There was a small heterogeneous hypersignal T1/mixed hyper-hyposignal T2 lesion with a “popcorn” like appearance, which was markedly hypointense on T2* weighted sequences, compatible with old hemosiderin deposits and various other blood products at different stages of evolution, at the right high frontal lobe, most likely resembling a cavernoma.

Cerebral angiography demonstrated multifocal DAVs along the SSS, supplied by bilateral middle meningeal arteries (MMAs), bilateral falcine arteries from the ophthalmic arteries (OAs), left meningohipophyseal trunk from the left internal carotid artery (LICA) and transosseous supply from bilateral superficial temporal arteries (STAs). There was drainage through the bilateral transverse sinus, medial occipital and left marginal sinus to bilateral jugular bulbs, which were moderately stenosed. Reflux into the cortical veins at the right frontoparietal lobe was noted. There was a small pial shunt from branches of the right ACA that likewise drained into the cortical veins at the right frontoparietal lobe.

Markedly delayed venous phase of the right cerebral hemisphere was observed with incomplete cavernous capture. Good cavernous capture on the left side was observed. The presumed MR diagnosis of DVA was confirmed only during the second angiography after partial embolization and reduction of the shunt flow, while during the first one, no DVA could be visualized.

The DVA drained the subcortical region of the right frontoparietal lobe through the transmedullary veins to a right medial cerebral vein, further draining into the vein of Galen, straight sinus and torcular respectively. Drainage of the DAVs into the DVA was also noted.

The diagnosis of infantile dural arteriovenous shunts was confirmed and the patient was treated by multiple transarterial glue embolizations using NBCA (n-butyl cyano-acrylate) into bilateral MMAs in order to decrease the shunt flow and thus the constraint to the brain

and drainage of the DVA for the patient to stay asymptomatic.

After the two embolization sessions, both hospitalizations were uneventful and the patient was discharged home remaining asymptomatic.

The patient is scheduled for follow-up MRI with further partial transarterial embolization in six months. If she develops any neurological symptoms or progressive venous congestion of the brain, then a more aggressive treatment (such as a transvenous approach) may be considered.

Discussion

Pediatric dural arteriovenous shunts (PDAVs)

While adult type dural arteriovenous shunts (DAVs) are defined as abnormal connections between the arteries and small venules within the wall of the sinuses or veins^{11,12}, mostly related to sinus thrombosis, venous hypertension or occurring after trauma or surgery^{9,13,14}, the pathogenesis of DAVs in children remains unclear. They account for approximately 10% of all intracranial arteriovenous shunts in children^{2,4,5}. Three groups of disease types can be identified, including dural sinus malformations (DSM) usually presenting in fetuses or neonates as a giant partially thrombosed epidural lake with moderate congestive cardiac failure (CCF), infantile DAVs which are multifocal and tend to present later, as in our case, and adult type DAVs of the cavernous plexus or sigmoid sinus¹⁻⁷.

Infantile DAVs are the most frequent type of pediatric DAVs reported in the literature^{2,3,5,7,15}. They are characterized by multifocal shunts with high-flow, low-pressure of the sinuses preserving the patency of the sinuses for long periods of time. Additionally induced pial shunts are frequently present¹. The most common presentation in neonates and infants is congestive cardiac failure (CCF), while older children can present with hydrodynamic disorders (hydrocephalus, macrocrania), developmental delay and seizures^{3,5}.

The neurological symptoms could be either related to venous congestion without reflux, leading to seizures, transient neurological deficit and mental retardation, depending on the degree of cavernous capture or pial reflux with venous hemorrhage, seizures and progressive neurological deficits¹. There are two types

Figure 1 A) Axial T2W MRI revealing a large tubular structure inferior to the corpus callosum, draining multiple dilated transmedullary veins at the right frontoparietal lobe to the vein of Galen. B) Coronal T2W and Sagittal T1W MRI demonstrating a small heterogeneous hypersignal T1/mixed hyper-hyposignal T2 lesion with a "popcorn" like appearance, compatible with old hemosiderin deposits, at the right high frontal lobe, most likely a cavernoma.

of progression of the disease, one is persistence of the high flow with pial induced shunts and the other is progressive outlet restriction, during skull base development with jugular stenosis or even occlusion^{1,4}, both of which were present in our case.

Developmental venous anomalies (DVAs)

Developmental venous anomalies (DVAs) are extreme limits of variability of the transcerebral venous system, leading to reduced flexibility that may result in venous ischemia or

Figure 2 A) Initial RECA angiograms lateral and AP views in early and late phase revealing DAVSs along the SSS, supplied by the right MMA, right STA, draining into the SSS, bilateral transverse-sigmoid sinuses to bilateral jugular bulbs and through the right medial occipital and marginal sinuses. There is stenosis of bilateral jugular bulbs with reflux into the cortical veins at the right frontoparietal lobe. B) Initial RICA angiogram lateral view in arterial and venous phases demonstrating supply of the right falcine artery to the AV shunt. A small pial induced shunt from branches of the branches of the right ACA draining into cortical veins at the right frontoparietal lobe is also noted. There is a markedly delayed venous phase of the right cerebral hemisphere with incomplete cavernous capture and poor visualization of the DVA.

infarctions¹⁶⁻²¹. In some cases, focal venous stenosis of the collecting vein at the point where it penetrates the dura to enter the dural sinus may be observed, leading to increased pressure within the DVA and precipitating thrombosis^{19,22}.

They are believed to be adaptations to accidents occurring during embryogenesis, between Paget's fourth to seventh stage (40-80 mm), re-

sulting in occlusion or maldevelopment of the superficial or deep veins. Due to plasticity of the vascular system at this stage, DVAs are formed as compensatory pathways^{23,24} recruiting and dilating preexisting transmedullary veins. The deep and superficial type of DVAs depends on the location of the venous collectors^{1,16}.

In our case, the DVA was of a deep type, col-

Figure 3 A) RICA and RECA angiograms 2nd embolization session in lateral view, venous phases revealing the DVA draining the subcortical region of the right frontoparietal lobe and the DAVSs through the transmedullary veins to the medial cerebral venous collector, further draining into the vein of Galen, straight sinus and torcular, respectively. B) 3D rotational RICA angiogram revealing the complex network of the dilated transmedullary veins, draining the right septal and thalamo-striate territories into a medial cerebral venous collector.

lecting the subcortical territories of the right frontoparietal lobe and transcallosal regions, including territories of the right septal and thalamostriate veins, draining into a large medial cerebral venous collector further draining into the vein of Galen and straight sinus, respectively. The course of a normal internal cerebral vein was not demonstrated. Incomplete cavernous capture of the right cerebral hemi-

sphere reflects the poor development of the superficial cortical veins.

Interestingly, the DVA was not well seen during the first angiogram, which was most likely due to the relatively high pressure within the torcular and transverse sinuses from the AV shunting along the SSS. Visualization of the DVA after partial embolization confirms that although there was no demonstrable retro-

grade flow into the straight sinus, there is definite congestion from the increased venous pressure^{1,8}. This also confirms the possibility of the DAVs to cause neurological symptoms in pediatric patients even though retrograde leptomeningeal drainage is not always identified¹.

Only a few cases of DAVFs associated with DVAs have been reported in the literature^{5,8-10}. Kuncz et Al¹⁰ reported a case in which the simultaneous presence of a DAVF and DVA in the same region caused venous infarction of the left frontal lobe, even though the DAVF did not drain directly into the DVA. As in cases of DVAs associated with arteriovenous malformations (AVMs), the DVA and its drainage route should be preserved, regardless of the type of treatment given²⁵⁻²⁷.

Conditions associated with DVAs

Our patient had a small cavernoma in the right frontal lobe within the territory of the DVA. Two forms of cavernomas have been reported, the inherited and sporadic types. To date, three gene mutations have been reported to be associated with the inherited or familial type including CCM1 (Chromosome 7q11.2-21 – Krit 1 gene), CCM2 (Chromosome 7p13-15 – MGC4607, malcavernin gene) and CCM3 (Chromosome 3q25.2-27 – PDCD10 gene)²⁸⁻³⁰. Since there was no family history of cavernomas and only a single lesion was detected, we presume it to be of a sporadic type.

Cavernomas have been reported to co-exist with DVAs in up to 33%^{21,31-34} and tend to develop within the territory drained by the DVA. There have also been multiple case reports of de novo formation of cavernomas after radiation, following sinus thrombosis and in DAVs with malignant cortical reflux³⁵⁻⁴⁰. The particular hemodynamic conditions created by a DVA and other causes of increased venous pressure could trigger an underlying vascular or abluminal defect that will be revealed as a cavernoma over time.

On the other hand, the association of DVAs with cortical dysplasias points to a developmental defect of the vasculature in a confined

region of the brain. Other types of malformations have been reported in association with DVAs including maxillofacial venous and lymphatic malformations, sinus pericranii, schizencephaly and multiple mucocutaneous venous malformations^{1,8,34,41-46}. There is as yet no evidence that these malformations and the DVAs are genetically related, however the DVA can be considered a time marker for the associated malformation¹. Concerning the history of patient's siblings with various types of malformations and the strong history of interfamilial marriage, there could be possible genetic mutations and chromosomal defects within the patient's generation.

Treatment

There were several reasons why urgent treatment was indicated in our patient even though she was asymptomatic. The presence of a DVA and its reduced capacity for alternations of the venous pressure and increased risk of symptoms including hemorrhage and venous infarctions, together with the evidence of congestion of the right cerebral hemisphere, cortical reflux, bilateral jugular bulb stenosis, pial induced shunt and multifocality which are poor prognostic factors support more active rather than conservative management¹⁻⁴.

The goal of the treatment for each type of DAVF is difficult to establish, as the patency of the sinuses should be preserved¹. As in this particular case, the drainage pathway of the DVA into the straight sinus and torcular, along with the congestion of the right cerebral hemisphere from incomplete cavernous capture suggests a high risk of sacrificing the SSS, therefore partial transarterial embolization was done. The aim was to reduce the shunt flow as much as possible for the child, who was asymptomatic, to develop normally. If any symptoms occur, either due to increased congestion from progression of the jugular bulb stenosis or new pial induced shunts, then more aggressive treatment may be needed. However, regardless of the type of treatment given, the DVA and its drainage pathway must be preserved.

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S. Geibprasert, M.D.
Diagnostic and Therapeutic
Neuroradiology Service
Hôpital Bicêtre, Le Kremlin-Bicêtre
Paris, France