Pediatric dural arteriovenous malformations

Report of 3 cases


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Pediatric dural arteriovenous malformations (dAVMs) are rare lesions that have a high mortality rate and require complex management. The authors report 3 cases of pediatric dAVMs that presented with macrocrania and extracranial venous distension. Dural sinus thrombosis developed in 2 of the cases prior to any intervention, which is an unusual occurrence for this particular disease. All 3 cases were treated using staged endovascular embolization with a favorable outcome in 1 case and a poor outcome in the other 2 cases. Complications developed in all cases and included dural sinus thrombosis, parenchymal hemorrhage, intracranial venous hypertension, and seizures. The strategies and challenges used in managing these patients will be presented and discussed, along with a review of the literature. While outcomes remain poor, the authors conclude that prompt treatment with endovascular embolization provides the best results for children with these lesions. A well-established venous collateral circulation draining directly to the internal jugular veins may further improve the rate of favorable outcome after embolization. (http://thejns.org/doi/abs/10.3171/2014.4.PEDS13695)

Key Words • dural arteriovenous malformation • sinus thrombosis • vascular disorders • endovascular embolization

Pediatric dural arteriovenous malformations (dAVMs) are extremely rare, with only 59 cases reported in the literature as of 2010. In a review by Barbosa et al. of 620 cases of intracranial pediatric AVMs reported from 1985 to 2003, only 52 (8.4%) were dAVMs, with the infantile subtype as the most frequent subtype. Pediatric dAVMs tend to be multifocal, with the transverse and sigmoid sinuses the most common sites of involvement. Pediatric dAVMs have a more aggressive clinical course relative to adult dAVMs, with a reported mortality rate greater than 25%. In this paper we report 3 cases of pediatric dAVMs that involved the transverse sinus associated with anomalous cerebral venous drainage, and describe their common presentation and clinical features.

Case Reports

Case 1

History and Examination. A 14-month-old boy presented with macrocrania and a 4-week history of progressive facial venous distension. His antenatal history was remarkable for preterm delivery at 36 weeks gestation. At presentation, his head circumference was tracking along the 97th percentile. Marked dilation of the facial veins was noted bilaterally. Neurological and developmental examinations were normal, and cardiology, ophthalmology, and hematology assessments were unremarkable.

Magnetic resonance imaging of the brain revealed a complex dAVM involving the right mastoid and petrous apex regions (Fig. 1A). The dAVM was fed by branches of the right middle meningeal artery (MMA) and the occipital and posterior auricular branches of the right external carotid artery (ECA). Cerebral venous outflow was anomalous, with complete occlusion of the sigmoid sinus–internal jugular vein (IJV) junction on the right and significant narrowing on the left. Venous drainage thereby occurred via enlarged cortical veins over the right hemisphere and thus into the right superior and inferior ophthalmic veins. Direct arterial fistulas from the right ECA to the right sigmoid sinus and cavernous sinus were also demonstrated. Subsequent digital subtraction angiography (DSA) confirmed the MRI findings and demonstrated retrograde flow along the right sigmoid and transverse sinuses (Fig. 1B).

First Endovascular Embolization and Postoperative Course. While under investigation the patient re-presented with focal seizures and tonic eye deviation, and 2 weeks...
later with irritability and vomiting following an unwit-
nessed fall with head strike. Repeat imaging studies re-
vealed acute thrombosis of the left sigmoid sinus extending
to the transverse sinus. Given the significant venous hy-
pertension induced by the thrombosis on a compromised
venous outflow, the patient underwent emergency endovas-
cular embolization of the right MMA and occipital artery
(OA) feeders of the dAVM, with a combination of the em-
bolic agent Onyx (ev3 Neurovascular) and polyvinyl alco-
hol. Anticoagulation with enoxaparin (Sanofi) was begun
and continued for 3 months to prevent further thrombosis
and to encourage recanalization of the left sigmoid sinus.
His right intraocular pressure (IOP) at the time was noted
to be elevated and was managed with latanoprost (Pfizer).

Over the next 6 months, repeat MRI studies dem-
onstrated decreased flow in the dAVM. The left sigmoid
sinus thrombosis initially recanalized, but later rethrom-
bosed with extension to the transverse sinus and torcu-
lar herophili. The right sigmoid sinus became partially
occluded and venous collaterals (dural and extracranial)
draining to the IJVs bilaterally were noted. These changes
were presumed to be related to altered vascular dynamics
induced by the endovascular embolization (Fig. IC).

Second Endovascular Embolization and Postopera-
tive Course. Two years after embolization, repeat DSA
demonstrated occlusion of the left transverse sinus, with
venous drainage now maintained by the cavernous sinus
and ophthalmic veins. New feeders to the dAVM from
the right OA and meningeal branch of the right intern-
al carotid artery (ICA) were observed. These findings,
along with increasing IOP despite treatment and a rap-
idly enlarging head circumference, prompted additional
intervention. He therefore underwent embolization of the
right OA and the meningeal branches of the right ICA
using Onyx. Anticoagulation was begun at this time for a
duration of 2 weeks after embolization. His postoperative
course was uneventful. At last review, 12 months after
his last embolization, he was clinically well with reduced
head circumference, IOP, and facial venous distension.
He has mild developmental delay.

Case 2

History and Examination. A 23-month-old boy pre-
sented with macrocrania and a 4-week history of right-
sided pulsatile proptosis on a medical history background
of recent viral infection associated with dehydration. His
antenatal history was unremarkable. Clinical examination
was significant for the above finding with a baseline head
circumference tracking along the 97th percentile. There
were no other ophthalmological findings. Neurological and
developmental examinations were normal and cardiology
and hematology assessments were unremarkable.

A brain MR image revealed a complex dAVM with
bilateral arterial feeders from the ECA (posterior auricu-

Fig. 1. Case 1. A: Axial (left), sagittal (center), and coronal (right) T1-weighted MR images after contrast administration show a dAVM at the right petrous apex with venous engorgement of cortical veins. B: Right common carotid angiogram showing a dAVM (arrow) draining to the right transverse sinus and demonstrating retrograde flow of the superior sagittal sinus. C: Axial T1-weighted postcontrast MR image showing left transverse sinus thrombosis.
lar and occipital arteries) and posterior cerebral artery, which was subsequently confirmed on DSA. Fistulous arterial feeders were located adjacent to the left transverse sinus and drained directly into it at the level of the torcular herophili. Normal venous outflow was significantly impaired due to subacute occlusive thrombosis of the left sigmoid sinus, superimposed on presumed congenital bilateral sigmoid sinus outflow stenoses. The majority of venous outflow therefore occurred in a retrograde manner via the superior sagittal sinus and cortical veins bilaterally, eventually draining into the right cavernous sinus (Fig. 2A and B).

**First Endovascular Embolization and Postoperative Course.** In view of his poor venous drainage and progressive proptosis, endovascular therapy was recommended to reduce arteriovenous shunting. Embolization of the left ECA feeders was performed using the embolic agent Onyx. Anticoagulation was not believed to be indicated due to the subacute nature of the sigmoid sinus thrombosis and the potential for hemorrhage, given the significant venous hypertension. His postoperative course was uneventful. At the 4-week review the child was clinically well with significant improvement of the proptosis. Decreased flow of the dAVM was noted on MRI.

Six months later he re-presented with complex partial seizures. A CT scan of his brain with contrast administration showed thrombosis of the left cortical veins and acute right sigmoid sinus thrombosis extending to the torcular herophili. Further endovascular treatment was not deemed appropriate at this stage due to the complexity of the dAVM and the presumptive risk of aggravation of the sinus thrombosis. He was started on anticoagulation therapy using enoxaparin in an attempt to recanalize the thromboses to improve venous outflow, which was partially successful.

**Subsequent Endovascular Embolizations and Postoperative Course.** Over the next 30 months, his seizure disorder increased in severity, with several admissions for status epilepticus. He developed unprovoked cortical venous thromboses. Radiographic studies during this time demonstrated chronic venous ischemic changes with atrophy of the left temporoparietal region and bilateral hippocampal sclerosis (Fig. 2C). These changes were associated with developmental, cognitive, and behavioral decline. Repeat DSA revealed new arterial feeders contributing to the dAVM at the level of the torcular herophili, originating from the posterior cerebral artery and temporal and occipital branches of the ECA bilaterally. Although he underwent 2 further embolization procedures to reduce the inflow to his venous system, these procedures were complicated by seizures, extensive venous sinus thromboses, and ventriculomegaly, despite the use of transient anticoagulation therapy. The last embolization was complicated by delayed intraparenchymal hemorrhage with intraventricular extension. In the setting of significant neurological decline with intractable seizures and poor

![Fig. 2. Case 2. A: Axial T2-weighted MR images show occlusive thrombus of the left sigmoid sinus (left) and early thrombosis of the right sigmoid sinus (center). An MR angiogram (right) shows a dAVM (arrow) adjacent to the left transverse sinus. B: Left common carotid angiogram showing a dAVM arising from branches of the left ECA and ICA. C: Axial T1-weighted postcontrast MR image (left) and T2-weighted MR image (right) show left temporoparietal chronic venous ischemic changes, enlarged cortical veins, and dilated torcular herophili.](image-url)
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conscious state, the patient’s family opted for palliative care, with subsequent death of the child 4 days later.

Case 3

History and Examination. This 3-year-old girl presented with macrocrania and gross-motor delay. Her antenatal history was significant for maternal systemic lupus erythematosus, for which her mother received azathioprine, hydroxychloroquine, prednisolone, aspirin, and enoxaparin throughout the pregnancy. At presentation the patient’s head circumference was tracking along the 97th percentile. Distended scalp veins were noted. She had gross-motor developmental delay. Her neurological examination was remarkable for hypertonicity, hyperreflexia, and nonsustained clonus bilaterally. Ophthalmological assessment revealed bilateral papilledema. Cardiology and hematology assessments were unremarkable.

A brain MR image demonstrated a dAVM in the left posterior fossa, adjacent to the torcular herophili. A Type I Chiari malformation was also demonstrated. Of note, there was no syringomyelia (Fig. 3A). Subsequent DSA confirmed the MRI findings and demonstrated bilateral feeders from the OA and posterior communicating arteries, with additional feeders from the right superficial temporal and vertebral arteries. Venous drainage occurred via a dominant right transverse sinus and IJV, and retrogradely via the straight sinus and superficial and deep cortical veins. There was reduced venous outflow via the left sigmoid sinus and IJV, secondary to narrowing of the left sigmoid sinus–IJV junction (Fig. 3B).

Endovascular Embolizations and Postoperative Course. In view of her clinical status, the patient underwent 2 endovascular embolization procedures over the next 2 months to reduce arteriovenous shunting. The left and subsequently right OA and MMA feeders were embolized using Onyx. Her postoperative course was uncomplicated in both instances. Follow-up brain MRI showed decreased flow in the dAVM.

One month after embolization, she presented with a transient episode of vomiting and lethargy that resolved spontaneously over 24 hours. Repeat imaging demonstrated no evidence of hemorrhage or dural sinus thrombosis. She re-presented the following month with sudden-onset vomiting, bradycardia, and obtundation. Imaging revealed an occlusive right-sided thrombus extending from the sigmoid sinus to the IJV, resulting in acute hydrocephalus and requiring emergency extraventricular drain insertion (Fig. 3C). In retrospect, her initial presentation after embolization may have been due to altered vascular dynamics with sluggish flow within the right transverse sinus. She was started on heparin; however, 2 weeks later, she developed significant intraparenchymal hemorrhage.

![Fig. 3. Case 3. A: Axial T1-weighted postcontrast MR image showing a dAVM in the left posterior fossa, adjacent to the torcular herophili. Dilated cortical veins are noted. B: Angiograms of the right ICA (left) and right ECA (center) show a dAVM nidus fed by branches of the right ICA and ECA. Anteroposterior view of a right ECA angiogram (right) demonstrates retrograde flow and narrowing of the left sigmoid sinus. C: Sagittal T1-weighted postcontrast MR image shows occlusive thrombosis of the right sigmoid sinus (arrow). D: Axial T2-weighted MR image (left) shows a large intraparenchymal hemorrhage associated with cerebral edema in the bilateral frontal lobes and basal ganglia. An axial T1-weighted postcontrast MR image (right) shows extension of the right transverse sinus thrombosis (arrow).](image-url)
with bilateral intraventricular extension, leading to immediate cessation of anticoagulation therapy and insertion of a new extraventricular drain.

Over the next 2 months, seizures and multifocal intraparenchymal hemorrhages contributed to the patient’s declining neurological status despite additional drainage attempts. Repeat MRI during this time showed evolution of the existing hematomas, progression of the thrombosis into the right transverse sinus, and diffuse cerebral edema in the frontal, temporal, and parietal regions bilaterally. Early signs of cortical liquefaction consistent with subacute ischemia were also present (Fig. 3D). Despite the inherent risk, anticoagulation was re-trialed in an attempt to restore venous outflow. Her neurological status, however, did not improve and her family elected for palliative care, with subsequent death of the child 5 days later.

Discussion

Pediatric dAVMs are commonly attributed to a disorder of venous sinus formation, with sinus wall overgrowth and abnormal venous space development in the sigmoid and transverse sinuses and torcular herophili.10,11,14,18 The embryology of cerebral venous development as described by Padget,5,8,10,23 Okudera et al.,17 and Raybaud15 can be divided into 7 stages.19,20,23 In stage 7A (week 12 of gestation), the conglomerate of channels that constitute the tentorial plexus of the primitive cerebrum merge to form the torcular herophili.19,20,23 This posterior dural plexus persists after birth. The increase in venous flow from the rapidly growing cerebral hemispheres leads to ballooning of the transverse sinuses between the fourth and sixth months of gestation. In pediatric dAVMs, it is believed that expansion and remodeling of the posterior sinuses persist beyond this time frame.3,6,8,11,14,18,23 In contrast, most cases of adult dAVMs are believed to be caused by thrombosis of a dural venous sinus, typically secondary to head trauma or a hypercoagulable state. The resulting occlusion and venous hypertension causes dilation of capillaries, which open direct shunts between dural arteries and veins.4,17,23 We believe that the dAVMs in all 3 of our cases developed from abnormal venous sinus formation, as the lesions were already well established on initial investigation and there was no evidence of a hypercoagulable state or significant head trauma prior to presentation. Regardless of the actual mechanism, the key factor in the development of adult or pediatric dAVMs appears to be decreased blood flow and subsequent occlusion and shunting within the dural venous sinus.4,5 The presence of anomalous sigmoid sinus–IJV outflow noted in all our cases would support this hypothesis.

Pediatric dAVMs have been classified into 3 types: dural sinus malformations, infantile dural arteriovenous shunts (dAVSs), and adult-type dAVSs.30,31,13 Dural sinus malformations are further divided into two subtypes: 1) those involving the posterior sinuses with or without the torcular herophili, with giant dural lakes and slow-flow mural arteriovenous shunting; and 2) those involving the jugular bulb with otherwise normal sinuses, but associated with a high-flow sigmoid sinus dAVM. Patients with dural sinus malformations typically present with high-output cardiac failure, venous sinus thrombosis, and consumptive coagulopathy in the antenatal or neonatal period.3,10,11,13 Infantile dAVSs are high-flow, low-pressure systems with patent sinuses and no venous lakes. Meningeal and occipital vessels are often directly involved. Clinically, infantile dAVSs manifest in early childhood with macrocrania, seizures, hydrocephalus, and progressive neurological deficits.7,11,13 Adult dAVSs present in older children and are nearly always located in the cavernous sinus. These are generally secondary to trauma, venous sinus thrombosis, or surgery, and present with venous hypertension, proptosis, and chemosis.7,9,10,13 Patients may also present with significant intracranial hemorrhage and neurological deficits if cortical venous hypertension is present.4 The arterial supply, venous drainage, sites of the dAVM, and clinical presentation of the cases in this series were similar to those reported in the literature with respect to infantile dAVSs. Interestingly, Cases 1 and 2 developed dural sinus thrombosis in early childhood prior to any intervention, which is atypical for this subtype of dAVM. We postulate that the thrombosis was precipitated by dehydration due to vomiting after concussion in Case 1, and a viral illness in Case 2.

Treatment of pediatric dAVMs is primarily via endovascular embolization, both transarterial and transvenous. Transumbilical embolization has been rarely used. Embolization has often been performed in stages to achieve gradual closure of the shunt in an attempt to maintain the patency of the abnormally enlarged dural sinus, allowing it time to undergo remodelling.6,8–11,13–15 Medical treatment with anticoagulants, diuretics, and inotropic agents have been used in this process to prevent dural sinus thrombosis and hyperdynamic cardiac failure.9,14 Radiation therapy has been used to treat adult dAVMs, but its role in pediatric dAVMs is less clear due to the young ages of the patients.9,12,14,15 Surgical treatment alone without previous embolization carries the risk of significant intraoperative blood loss with high rates of mortality and morbidity.6,9,10,13–15 Direct surgical intervention has only rarely been reported, such as by Monges et al.16 Our initial management decisions in Cases 1 and 2 were affected by the coexisting venous sinus thromboses, due to the significant risk of causing further thrombosis and obstruction of an already compromised drainage system. While clinical and radiological improvement was achieved in the immediate postoperative period using transarterial embolization, additional thromboses did eventually develop in both cases. The management of pediatric dAVMs in the context of coexisting dural sinus thrombosis is poorly documented due to the rarity of these lesions.

Outcomes of treatment for pediatric dAVMs are poor. Morita et al.17 reported an overall mortality rate of 38% in a literature review up to 1995. This was consistent with the findings of Barbossa et al.1 who reported a mortality rate of 37.9% in 2003. As of 2010, the overall rate has dropped to 26%,10 but it is unclear if this was due to better embolization techniques or a lack of follow-up in these cases. Dural sinus malformations are reported to be associated with a better prognosis when there is unilateral dural sinus involvement remote from the torcular herophili or superior sagittal sinus, due to adequate al-

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ternate cerebral venous drainage in the event of involved sinus thrombosis or occlusion. The presence of bilateral cavernous sinus drainage and the absence of jugular bulb tympanus or occlusion are also favorable factors in the prognosis of dural sinus malformations.1,6,14 We are unaware of any previous reports in the literature that suggest these factors apply to infantile dAVMs, but we note that while the torcular herophili was involved only marginally in Case 1, it was extensively involved in the other cases as the site of the dAVM. Venous outflow obstruction secondary to occlusive dural sinus thrombosis was present in all cases, but only Case 1 developed collaterals draining directly to the IJVs. These factors may have accounted for the favorable outcome of Case 1.

Transarterial embolization often results in the recruitment of secondary arterial feeders, which may lead to a recurrence of symptoms. This was consistent with our series, in which Cases 1 and 2 developed significant recruitment subsequent to endovascular therapy. Excessive tortuosity of the affected vessels has also been reported to hamper attempts at transarterial embolization.5,9,11,15 Transvenous embolization is typically only used when there is sufficient venous collateral circulation, as it usually results in complete occlusion of the affected sinus. The rerouting of blood through cortical veins and deep venous sinuses to ultimately drain into the superior ophthalmic and facial veins has been described previously in the literature and was well demonstrated in our series.2,9,11,15

All 3 of our cases experienced aggravation or formation of dural sinus thrombosis within 9 months of embolization, which resulted in increased venous hypertension. This is turn led to progressively increasing IOP in Case 1 and complex seizure disorders in Cases 2 and 3. Additionally, Cases 2 and 3 were further complicated by the development of intracerebral hemorrhages likely secondary to antiocoagulation therapy. While antiocoagulation therapy for dural sinus thrombosis improves outcomes even in the presence of intracranial hemorrhage,21 the guidelines for management of these lesions in the context of coexisting infantile dAVMs are unclear.

Conclusions

In this paper we report 3 cases of pediatric dAVMs in children illustrating the pathogenesis, clinical subtypes, and current management of this rare disease. We also note the atypical development of dural sinus thrombosis in 2 of our cases prior to intervention, which made subsequent management significantly more challenging. While outcomes remain poor for children with these lesions, prompt treatment with endovascular embolization provides the best results.5,6,10,14 Our experience suggests that a well-established venous collateral circulation draining directly to the IJV may further improve the rate of favorable outcome after embolization.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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