Endovascular correction of an infantile intracranial venous outflow obstruction

Case report

MARYAM SOLTANOLKOTABI, M.D.,1 SHAHRAM RAHIMI, M.D.,1 MICHAEL C. HURLEY, M.D.,1,2 ROBIN M. BOWMAN, M.D.,2 ERIC J. RUSSELL, M.D.,1 SAMEER A. ANSARI, M.D., PH.D.,1,3 AND ALI SHAIBANI, M.D.1,3

Departments of 1Radiology and 2Neurosurgery, Northwestern University Feinberg School of Medicine; and 3Department of Medical Imaging, Children’s Memorial Hospital, Chicago, Illinois

The authors report on the case of a 7-year-old boy who presented with a reduced level of activity, macrocephaly, prominent scalp veins, and decreased left-sided visual acuity. Imaging workup demonstrated generalized cerebral volume loss, bilateral chronic subdural hematomas, absent left sigmoid sinus, hypoplastic left transverse sinus, and severe focal weblike stenosis of the right sigmoid sinus. Right sigmoid sinus angioplasty and stent insertion was performed, with an immediate reduction in the transduced intracranial venous pressure gradient across the stenosis (from 22 to 3 mm Hg). Postprocedural diminution of prominent scalp and forehead veins and spinal venous collateral vessels was followed by a progressive improvement in visual acuity and physical activity over a 1-year follow-up period, supporting the efficacy of angioplasty and stent insertion in intracranial venous outflow obstruction.

There are multiple potential causes of intracranial venous hypertension in children. Development of dural sinus stenosis in infancy may be one such cause, mimicking the clinical presentation of other causes such as vein of Galen malformations. This condition can be ameliorated by early endovascular revascularization.

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vascular disorders  
i intracranial sigmoid sinus stenosis  
i intracranial venous outflow obstruction  
macrocephaly

The dural venous sinuses are the primary channels for drainage of intracranial blood and CSF resorption and, unless collateral pathways offer an adequate alternative drainage route, a stenosis or obstruction may result in venous hypertension followed by elevated intracranial and CSF pressure.3 Stent placement can reduce the pressure gradient across a stenosis, improve venous drainage, and reduce or eliminate clinical symptoms, as occurred in this case.6

**Case Report**

History and Examination. This 7-year-old boy presented after an ophthalmological examination for decreased visual acuity demonstrated bilateral optic pallor, and a subsequent CT scan showed bilateral subdural hematomas (SDHs). His parents thought that he had always had poor stamina and was unusually tentative with physical activity. His examination demonstrated macrocephaly, prominent scalp veins, mild left exotropia, and bilateral optic nerve pallor, with 20/20 vision OD and 20/80 vision OS. He was born at term with intrauterine growth retardation, pulmonary stenosis requiring an urgent balloonoplasty, and congenital absence of the right kidney. He later underwent bilateral myringotomies for troublesome serous otitis media.

The patient’s head circumference had steadily increased from the 10th percentile at 9 months of age to the 95th percentile at 19 months, and an MR image of the brain obtained at another institution at that time showed generalized cerebral volume loss with mild posterior periventricular T2 hyperintensity of white matter and no obvious venous abnormality. A second MRI sequence of the brain and additional MR venogram at age 32 months additionally revealed, in retrospect, a focal severe stenosis of the
right sigmoid sinus, whereas the prepontine and premedullary veins appeared normal at that time.

Results of genetic screening by chromosomal microarray analysis and the Noonan syndrome panel (Harvard) were both negative. Results of hematological testing for coagulopathy were normal.

Neuroimaging Studies. Repeat imaging was performed after presentation to our institution. A CT head scan showed generalized cerebral volume loss with scattered frontal lobe calcifications (Fig. 1A) and bilateral SDHs. The MRI sequence demonstrated dilated optic nerve sheaths (Fig. 1B and C). A CT venography study demonstrated an absent left transverse sinus and severe focal weblike stenosis of the right transverse sinus, with cortical venous ectasia, enlarged venous channels paralleling the sagittal and right transverse sinuses, a persistent median prosencephalic vein with a patent straight sinus, and dilated pontomesencephalic and pontomedullary veins communicating with a tortuous and enlarged ventral longitudinal spinal vein (Fig. 2).

These findings were interpreted as consistent with intracranial venous hypertension with both superficial and deep collateral venous drainage. The generalized volume loss and calcification of the brain parenchyma were thought to be due to longstanding intracranial venous congestion.

Operation. After consultation with the patient’s neurosurgeon, a decision was made to proceed initially with bur-hole drainage of the larger, left-sided chronic SDH, to be followed by cerebral angiography and venography and with possible endovascular treatment 10 days later.

Cerebral angiography showed that the supratentorial venous drainage was via the stenotic right transverse/sigmoid sinus as well as collateral scalp and facial veins. Venous drainage of the posterior fossa was primarily via dilated prepontine and premedullary veins extending to the ventral longitudinal veins of the spinal cord, and from there to the cervical epidural venous plexus (Fig. 3). Venous pressures were measured in the dural venous sinuses proximal to the focal severe stenosis of the right sigmoid sinus as well as in the distal sigmoid sinus and jugular bulb. The venous pressure proximal to the stenosis was 23 mm Hg, with a pressure of 1 mm Hg distal to the stenosis (Fig. 4 left).

The sigmoid sinus proximal to the stenosis was measured at a maximum diameter of 10 mm, and an 8-mm, self-expanding, open-cell nitinol stent (Cook Zilver 8 × 20 mm) was positioned across the stenosis under venous roadmap guidance. A 6-mm angioplasty balloon was used for poststent dilation of a residual waist (Fig. 4 right).

Venous pressure measurements were repeated following stent placement, demonstrating a proximal (to the stent) sigmoid pressure of 7 mm Hg, with a right internal jugular vein bulb pressure of 1 mm Hg. Repeat angiography revealed an immediate decrease in the size of the dilated prepontine/premedullary veins. The patient was placed on a daily regimen of 40 mg of aspirin and was discharged on the 3rd morning postprocedure with no complications.

Postoperative Course. The patient was closely followed by the neurosurgical and neurointerventional teams.
nous channel in the superior sagittal sinus (arrow), which in turn drain into the cervical epidural plexus, prepontine veins extending to the longitudinal veins of the spinal cord, posterior fossa venous drainage primarily via dilated premedullary and sinus cephalic vein pressure gradient of 6 mm Hg.

At 1 month he had improved significantly. The facial and scalp veins had reduced in size and, according to his parents, he demonstrated increased physical activity and energy; they described him as a “different kid.” One-year follow-up demonstrated continued normal neurological development, with a very active boy who was able to participate normally in school and extracurricular activities. Ophthalmological evaluations showed a slight improvement in visual acuity on the left and stable vision on the right side.

Results of cross-sectional imaging of the brain remained stable at 3 months. Follow-up angiography, venous pressure measurements before stent placement showed a pressure of 23 mm Hg in the right sigmoid sinus (white arrow) and duplicated channel in the right sigmoid sinus (black arrow).

Fig. 3. Preprocedure angiograms. A: Persistent median prosencephalic vein (black arrow) and duplicated channel in the right sigmoid sinus (white arrow). B: Absence/occlusion of left sigmoid sinus (arrow). C: The venous phase of the vertebral angiogram demonstrates posterior fossa venous drainage primarily via dilated premedullary and prepontine veins extending to the longitudinal veins of the spinal cord, which in turn drain into the cervical epidural plexus. D: A parallel venous channel in the superior sagittal sinus (arrow).

Fig. 4. Left: Venous pressure measurements before stent placement showed a pressure of 23 mm Hg in the right sigmoid sinus proximal to the stenosis (asterisk) and 1 mm Hg in the right jugular bulb distal to the stenosis (dagger); the venous pressure gradient was 22 mm Hg. Right: Cook Zilver 8 × 20–mm stent placed in the right sigmoid sinus. Venous pressure following stent placement was 7 mm Hg proximal to the stent and 1 mm Hg distal to the stent, with a resultant pressure gradient of 6 mm Hg.

Discussion

Because of the common pathophysiology of venous hypertension, it is not surprising that this child with chronic intracranial venous outflow obstruction presented similarly to a typical non-neonatal patient with a vein of Galen malformation (VGM): increasing head circumference, prominent scalp veins, cerebral calcification, and volume loss. Most striking of all was the finding of a persistent median prosencephalic vein of Markowski (MPVM), which is pathognomonically associated with VGM. The lack of hydrocephalus in our case (a common occurrence with VGMs) is most likely related to the timing, mechanism, and degree of venous hypertension.

Vein of Galen malformations are congenital and therefore interfere with the normal maturation of CSF resorption pathways very early on. In the case presented, the venous outflow obstruction most likely occurred after birth (low-normal initial head circumference) but before the closure of either main fontanel (symmetrical enlargement), because the anterior fontanel usually closes between 4 and 24 months of age (90% between 7 and 19 months), whereas the posterior fontanel closes between 8 and 18 months of age. The median prosencephalic vein is thought to be an in utero venous channel that regresses and disappears during normal development, in parallel to development of the straight sinus. In patients with VGM the persistent MPVM and absent straight sinus are considered a consequence of fetal intracranial venous hypertension. The presence of an MPVM and its subsequent regression after resolution of the venous hypertension in this case is of interest. This finding may suggest that the MPVM could potentially persist as a venous outflow channel for some time after birth; however, this speculation should be interpreted with caution due to the absence of early vascular imaging.

In the case of our patient, once the focal severe web-like stenosis of the right sigmoid was identified as the likely cause of the problem, a multidisciplinary assessment identified endovascular treatment of the lesion as the primary approach. In case of technical failure, the probable alternative would have been CSF diversion by a ventriculoperitoneal or lumboperitoneal shunt, which would not have directly addressed the venous hypertension.

Selection of the stent was based on a balance between

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diameter, flexibility, and size of the delivery system. The normal size of the adult sigmoid sinus is approximately 10–12 mm. Stents in this size category are generally less flexible and require larger delivery sheaths than were thought to be safe to use in a 7-year-old child. An 8-mm stent was chosen as a good compromise and with good results.

Enlarging head size and evidence for collateral venous channels can occur with any cause of intracranial venous hypertension (and consequent elevated intracranial pressure) acquired prior to the closure of the fontanels and sutures. In the absence of a VGM or arteriovenous shunt, a detailed examination of the venous outflow of the brain is necessary. The patient described herein was successfully treated with the endovascular technique, with good short-term, 1-year, and 3-year results. Long-term follow-up of this patient is necessary to exclude future development of venous outflow insufficiency due to causes such as in-stent stenosis or inadequacy of the stent size.

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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