Hydrocephalus in a patient with an unruptured pial arteriovenous fistula: hydrodynamic considerations, endovascular treatment, and clinical course

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Intracranial pial arteriovenous fistulas, also known as nongalenic fistulas, are rare vascular malformations affecting predominantly the pediatric population. Hydrocephalus is an unusual presentation in which the exact pathophysiology is not fully understood. The aim of treatment in these cases is occlusion of the fistula prior to considering ventricular shunting. Here, the authors describe the hydrodynamic considerations of the paravascular pathway and the resolution of hydrocephalus with endovascular treatment of the fistula.

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Intracranial pial arteriovenous fistulas (AVFs), also known as nongalenic fistulas, are rare vascular malformations affecting predominantly the pediatric population. Prevalence remains unknown due to their rarity but is estimated to be between 0.1/100,000 and 1/100,000 from previous studies.23,44 The distinct angioarchitectural characteristics include a high-flow shunt of 1 or more feeding arteries to an early draining cortical vein without intervening nidus of a typical arteriovenous malformation. These are distinguished from a dural AVF by the location of the fistula site in the subpial meningeal space, and from a vein of Galen malformation, by the lack of direct involvement of the embryonic median prosencephalic vein.2,15,24,32,36,43

Clinical presentation depends on age, mass effect, hemodynamic steal, or venous hypertension. In the newborn a pial AVF usually presents with congestive heart failure, increasing head circumference, or focal neurological deficit.11,31 Adolescents and adults tend to present with headache, seizures, and focal neurological deficit. Acute presentation due to subarachnoid or intracerebral hemorrhage is less common.1,4,8,16,25,33,37,44

Hydrocephalus is an unusual presentation but can be seen in newborns and infants. The exact pathophysiology of this clinical presentation is still unknown but is likely due to the raised venous pressure caused by a high-flow fistula. The difference in the CSF dynamic between the child and the adult might have a significant role in the differing presentations based on age.9,12,29

In this article we describe the hydrodynamic considerations in the resolution of hydrocephalus with endovascular treatment of a pial fistula with transarterial embolization using coils and glue (N-butyl cyanoacrylate [NBCA]).

Case Report

A 4-year-old boy was referred to our hospital for a second medical opinion. He had a history of 2 years of severe cognitive developmental disturbances; 9 months of insidious, intermittent headache and irritability; and an isolated episode of tonic-clonic seizure lasting a few seconds with a postictal period of an hour. Psychomotor and gait disturbances were not noticed by the parents until their 2-year-old son began to surpass his older sibling in neurocognitive development.

The patient was treated with oxcarbazepine and was considered for CSF shunt surgery by another neurosurgeon, a procedure to which the parents did not consent.

On our examination the infant was noted to demonstrate remarkable psychomotor retardation, was unable to complete sentences, babbled on occasion, was unable to walk, demonstrated mild somnolence, exhibited bilateral papilledema, and had notable facial venous distention. There were no focal clinical findings in the remaining portion of the neurological examination.
CT scans from another institution demonstrated hydrocephalus, transependymal edema, juxtacortical calcifications, and subarachnoid dilated vessels in the sylvian sulcus and perimesencephalic cistern (Fig. 1). A CT angiogram of the head revealed an AVF supplied by a single branch of the left middle cerebral artery with a large intervening aneurysm draining to a cortical vein (Fig. 2).

The child underwent digital subtraction angiography (Fig. 3), which confirmed a pial AVF with an aneurysmal venous dilation between a single feeder, corresponding to...

FIG. 1. Unenhanced head CT scans obtained at another institution, showing dilated perimesencephalic (A) and sylvian (B) vessels, supratentorial hydrocephalus with transependymal edema (C), and bilateral and symmetrical hyperdense lesions (D) in the supratentorial juxtacortical areas.

FIG. 2. In the arterial phase, hyperdense, dilated tortuous arterial and venous vessels are shown. Left: The left middle cerebral artery is larger in diameter than the right middle cerebral artery. Right: In the rostral part of the left lateral sulcus, a saccular lesion communicates with a superficial cortical vein associated with dilation of the superficial and deep veins.

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FIG. 3. Catheter angiograms showing a pial AVF with an aneurysmal venous dilation between a single feeder, corresponding to a left M3 afferent artery, and a vein at the sylvian fissure (A), with high efferent drainage to the superior sagittal sinus and retrograde reflux of deep venous system from the confluence of sinuses to both cavernous sinuses (B) and also retrograde reflux of contralateral cerebral veins to petrosal sinus (C). Moreover, there were stenoses (hypoplasia) of both sigmoid sinuses and both jugular bulbs were formed by ipsilateral petrous sinuses. The fistula was excluded from the cerebral circulation by means of embolization of the venous pouch with coils and glue (NBCA, D–G).
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The child was discharged 3 days after embolization without complications. An MRI study and 3-month posttreatment CT angiogram (Fig. 4) demonstrated considerable reduction of hydrocephalus. The patient has recovered well, and his development thus far has been appropriate for his age as assessed by our neuropsychologist.

Discussion

Similar to vein of Galen malformations, relatively more common high-flow fistulas, the clinical presentation of pial AVF in pediatric patients tends to vary by age. Hydrocephalus and macrocrania have been reported to be between 14.3% and 38% in different clinical series.15,24,33,39 In patients with pial AVF and mild to moderate hydrocephalus without acute intracranial hypertension, the aim of treatment is occlusion of the fistula, either surgical21,40 or endovascular. Ventriculoperitoneal shunting prior to this exclusion decreases the pressure in the ventricles that may result in hygromas, subdural hemorrhage,2 or hemorrhage as shown in different clinical scenarios with venous overload and associated hydrocephalus.26,30,34,41 This paradigm shift is clearer in vein of Galen malformation treatment in which endovascular obliteration is the first-line option rather than direct management of the hydrocephalus by CSF diversion.10,11,27

Endovascular treatment has been reported in other articles to be effective in the obliteration of the fistula, with excellent outcomes in most patients.14,17,38,39,43 The technique is challenging because of the high-flow nature of the fistula and because distal migration of embolic agents through the shunt cannot be easily controlled; inadvertent migration of these agents into the veins may result in immediate hemorrhage by venous outflow obstruction.2,13,20,28

We decided to use NBCA in our patient in combination with coils because of our vast experience using this material.

Understanding of CSF physiology is evolving since the bulk flow model. In children, it is well established that communicating hydrocephalus can be caused by vascular dysfunction, such as idiopathic venous outflow resistance, venous sinus collapse, venous thrombosis, and venous outflow stenosis at the skull base associated with craniofacial dysostoses. The exact mechanism, like in the pial AVF, is unclear.5,22

In this case, the arteriovenous shunt was considered the cause of hydrocephalus, and therapeutic embolization was planned. We speculate that excessive intracranial venous pressure can affect the paravascular pathway secondary to an inverted hydrodynamic flow.2,29 This paravascular pathway may play a significant role as arachnoid granulations lack maturation.27,44 This paravascular pathway has already been shown in experimental models, but not in humans,5,18,19 as an alternative CSF passage and is believed to depend on hydrodynamics, if there is venous hypertension from venous overload, like in pial AVF, decreased CSF resorption will occur, leading to nonobstructive hydrocephalus.9,12

In this case, the dystrophic calcification shown in the images can be an accumulation of waste product in the paravascular space that cannot be washed out because of the inverse hydrostatic pressure29 and not only the result of chronic hypoxic insult described by others.9,15,27,29,35,42 This evolving understanding of CSF dynamics will help us bring better care and make better decisions, but many
animal models and clinical analyses on hydrocephalus will be needed to fully understand the hydrodynamics.

References

**Disclosures**

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

**Author Contributions**

Conception and design: Morales-Gómez. Acquisition of data: Morales-Gómez, Garza-Oyervides. Analysis and interpretation of data: Morales-Gómez, Mercado-Flores, Elizondo-Riojas, Boop, Martínez-Ponce de León. Drafting the article: Morales-Gómez, Mercado-Flores, Elizondo-Riojas. Critically revising the article: Morales-Gómez, Garza-Oyervides, Boop, Martínez-Ponce de León. Reviewed submitted version of manuscript: Morales-Gómez, Garza-Oyervides, Arenas-Ruiz, Boop, Martínez-Ponce de León. Approved the final version of the manuscript on behalf of all authors: Morales-Gómez. Administrative/technical/material support: Morales-Gómez, Martínez-Ponce de León. Study supervision: Morales-Gómez, Martínez-Ponce de León.

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