Hydrocephalus in unruptured brain arteriovenous malformations: pathomechanical considerations, therapeutic implications, and clinical course

Clinical article

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Objective. The goal in this study was to present possible pathological mechanisms, clinical and imaging findings, and to describe the management and outcome in patients with hydrocephalus due to unruptured pial brain arteriovenous malformations (AVMs).

Methods. Medical records and imaging findings in 8 consecutive patients with hydrocephalus caused by AVMs and treated between June 2000 and September 2007 were retrospectively reviewed to determine clinical symptoms, AVM location, venous drainage, level/cause of obstruction, and degree of hydrocephalus. Management of hydrocephalus, AVM treatment, complications, and follow-up results were evaluated.

Results. Headaches were the most common clinical symptom (7 of 8 patients). Deep venous drainage was identified in all patients. Mechanical obstruction by the draining vein or the AVM nidus was seen in 6 patients, in whom obstruction occurred at the interventricular foramen (2 patients) or the aqueduct (4 patients). Hydrodynamic disorders following venous outflow obstruction and venous congestion of the posterior fossa led to hydrocephalus in the remaining 2 patients. Ventriculoperitoneal (VP) shunts were placed in 6 of 8 patients with a moderate to severe degree of hydrocephalus. Regression of hydrocephalus was noted in 4 patients, whereas in 2 the imaging findings were stable, 1 of whom had decreased hydrocephalus only after AVM size reduction. In 2 patients with mild hydrocephalus who were not treated with shunt insertion, 1 improved and 1 was clinically stable after AVM treatment.

Conclusions. The most common cause of hydrocephalus in unruptured brain AVMs is mechanical obstruction by the draining vein if it is located in a strategic position. Management should be aimed at treatment of the AVM; however, VP shunts may be necessary in acute and severe cases of hydrocephalus. (DOI: 10.3171/2008.7.JNS0815)

Key Words • arteriovenous malformation • hydrocephalus • ventriculoperitoneal shunt

Pial brain AVMs are defined as abnormal conglomerates of dilated arteries and veins within the brain parenchyma without the normal intervening capillary bed, resulting in arteriovenous shunts. Although believed to be congenital lesions, they usually present later in life, usually around the 2nd or 3rd decades. The most common presentation of symptomatic brain AVMs is intracranial hemorrhage, which may be intraparenchymal, intraventricular, and/or subarachnoid in location. Other, less common presenting symptoms are epilepsy, nonspecific headaches, and focal neurological deficits. Symptoms of increased intracranial pressure with imaging signs of hydrocephalus are unusual in adults, and to the best of our knowledge, there have been only 5 case reports in the pertinent literature that described an angiographically detectable AVM as the source of hydrocephalus in adults. In the pediatric population, however, hydrocephalus may be the presenting symptom in up to 38% of cases.

We report on 6 adult and 2 pediatric patients with unruptured AVMs presenting with hydrocephalus. We discuss the possible pathological mechanisms, and, based on these, their respective management strategies; our goal is to raise the awareness of this possible presenting symptom and potential complication of pial brain AVMs.
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Methods

Following a retrospective analysis of cases recorded in the dedicated and prospectively entered neurovascular databanks of our hospitals between June 2000 and September 2007, we identified 8 from a total of 440 patients with brain AVMs seen during that period whose primary symptoms were related to hydrocephalus from unruptured brain AVMs. Pediatric patients presenting with vein of Galen AVMs and pial arteriovenous fistulas were not included. The medical records were reviewed for patient sex, age, and clinical symptoms. Imaging and angiographic findings were used to determine the angioarchitecture of the AVM, regarding its location, size, and venous drainage, the level and the cause of obstruction, and the degree of hydrocephalus. Management of the hydrocephalus (that is, whether a VP shunt placement procedure was performed), treatment of the AVM, complications, and follow-up results were documented. The study was approved by the local institutional review boards.

Results

The patient demographics are shown in Table 1. There were 4 male and 4 female patients, whose ages ranged from 2 to 55 years, with a mean age of 24.5 years. Headaches were the most common presenting symptom, being present in 7 of 8 cases, and they usually preceded the onset of seizures or other neurological deficits. In 1 pediatric patient, macrocrania was the first presenting symptom of hydrocephalus and led to further imaging workup. One patient had a cerebrofacial arteriovenous metameric syndrome, with AVMs at the left basal ganglia and left optic nerve, whereas the remaining patients had single, glomerular, nonfistulous AVMs. The location of the AVMs varied; however, the dominant venous drainage was directed into the deep venous system in all patients. The mean follow-up duration was 26 months (range 2–72 months).

The size of the AVM varied from 1.5 to 8 cm. Five AVMs were located supratentorially and 3 infratentorially, with all being of the parenchymal type. Single deep venous drainage was present in 2 of the 5 supratentorial AVMs, whereas the remaining 3 cases had deep venous drainage as the major pathway, with minimal drainage through small cortical veins. In all 3 patients with infratentorial AVMs, the lesion drained through multiple cerebellar veins, further draining into tributaries of the deep venous system.

Mechanical obstruction of the ventricles by a dilated drainage vein was identified in 4 cases, occurring at the foramen of Monro and at the level of the aqueduct in 2 cases each (Figs. 1 and 2). At the foramen of Monro, circumscribed ectasias of a dilated thalamostriate vein draining further into the ICVs were identified as the reason for the obstruction and resulted in unilateral lateral ventricular dilation in both cases (Fig. 3). At the level of the aqueduct, a venous pouch of ventricular vein and a dilated vein of Galen served as the major draining pathway, which led to pressure on the tectum and resulted in critical stenosis of the aqueduct. In 2 patients with AVMs located in the midbrain and cerebellum, respectively, the mechanical obstruction was caused by the nidus itself at the level of the aqueduct. In the remaining 2 patients with AVMs located in the cerebellum, there was no visible mechanical ventricular obstruction or stenosis. However, there was evidence of venous outflow obstruction with congestion of the posterior fossa and the deep venous system. This was due to occlusion of the straight sinus in 1 adult (Case 7, Fig. 4) and jugular bulb occlusion on one side with stenosis on the other side in a 2-year-old boy (Case 8).

Six of the 8 patients, all with a moderate to severe degree of hydrocephalus, underwent VP shunt placement performed using a Codman-Hakim programmable valve system, with initial pressure settings of 160 mm H2O. There was an unusually high amount of complications related to these shunts, despite normal CSF laboratory results: 2 patients had malfunctioning shunts caused by obstruction of the device, with worsening of the hydrocephalus leading to shunt revisions during the same hospital admission. Subsequently, both patients underwent 2 and 4 shunt revisions, respectively; all were deemed due to recurring obstruction of the intracranial shunt. In 3 patients, the following complications occurred: 2 patients had SDHs (1 of which recurred even after adjusting the pressure settings), 1 had hygromas (which did not recur after adjusting the pressure settings), and 2 had slit ventricles. The hydrocephalus remained unchanged in 2 patients without clinical worsening; in 1 it improved only after the radiosurgery had taken effect, resulting in decreased size of the AVM nidus. Of the 6 patients treated with a VP shunt, only 1 had clinical and imaging improvement after the shunt insertion procedure. In the 2 patients in whom no shunt was placed, 1 had improvement of the hydrocephalus after embolization of the AVM, and the other, who had only mild hydrocephalus, was clinically stable with conservative treatment.

Treatment of the AVM was performed in 6 patients, as follows: 1 was treated with radiosurgery alone; in 4 patients partial embolization with glue (N-butyl cyanoacrylate) was performed to reduce the shunt volume or to target angioarchitectural weak points, followed by radiosurgery; and the patient with the cerebrofacial arteriovenous metameric syndrome died after a prolonged status epilepticus occurred between scheduled treatment sessions. In 2 patients conservative management was adopted because of the eloquent location and large size of the AVM in one (midbrain location and involvement of both cerebellar hemispheres; Case 6) and the fact that the patient was too young to receive radiosurgery in the other (Case 8).

Discussion

Pathophysiological Features of Hydrocephalus Related to Brain AVMs

The most common cause of hydrocephalus in a patient with a pial brain AVM is the rupture of an intraventricular or subarachnoid hemorrhage, with subsequent blockage of the arachnoid villi or cisterns surrounding the
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Symptoms</th>
<th>Lesion Location</th>
<th>Lesion Size (cm)</th>
<th>Venous Drainage</th>
<th>Level &amp; Cause of Obstruction</th>
<th>Degree of HC</th>
<th>VP Shunt Op/Result</th>
<th>Shunt-Related Complications</th>
<th>AVM Tx/Result</th>
<th>Final Angiographic Result</th>
<th>FU (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>16, F</td>
<td>progressive HA for 7 mos</td>
<td>rt thalamus</td>
<td>1.5</td>
<td>lat perimesencephalic vein → VG</td>
<td>severe; impending herniation</td>
<td>yes/unchanged</td>
<td>none</td>
<td>embol/improved</td>
<td>small residual nidus (&lt;1 cm)</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>17, F</td>
<td>HA for 2 mos w/ sudden alteration of consciousness</td>
<td>lt basal ganglia &amp; thalamus</td>
<td>6</td>
<td>thalamostriate vein → lt ICV → VG</td>
<td>severe; midline shifting</td>
<td>yes/improved</td>
<td>bilat hygromas, slit ventricles</td>
<td>embol/initially stable, then died (status epilepticus)</td>
<td>2</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>35, M</td>
<td>HA</td>
<td>rt temporoparietal lobe</td>
<td>7</td>
<td>ventricular vein → thalamostriate vein → rt ICV → VG</td>
<td>severe</td>
<td>yes/unchanged</td>
<td>none</td>
<td>embol, then RS/ improved</td>
<td>complete obliteration</td>
<td>30</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>39, F</td>
<td>HA followed by seizures</td>
<td>rt frontal-callosum</td>
<td>3</td>
<td>thalamostriate vein → rt ICV → VG</td>
<td>mild</td>
<td>no</td>
<td>NA</td>
<td>embol, then RS/ improved</td>
<td>small residual nidus (&lt;1 cm)</td>
<td>34</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>6, F</td>
<td>seizures followed by HA &amp; papilledema</td>
<td>rt midbrain</td>
<td>2.5</td>
<td>lat perimesencephalic vein → VG</td>
<td>moderate</td>
<td>yes/improved</td>
<td>multiple revisions; bilat SDHs</td>
<td>RS/stable</td>
<td>small residual nidus (&lt;1 cm)</td>
<td>72</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>26, M</td>
<td>chemosis/ proptosis of rt eye, progressive HA</td>
<td>cerebellum</td>
<td>8</td>
<td>cerebellar veins → VG</td>
<td>severe; impending herniation</td>
<td>yes/improved</td>
<td>multiple revisions; bilat SDHs; slit ventricles</td>
<td>none/stable</td>
<td>unchanged nidus (8 cm)</td>
<td>36</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>55, M</td>
<td>progressive HA for 3 mos</td>
<td>lt cerebellum</td>
<td>3</td>
<td>cerebellar veins w/ severe PF congestion</td>
<td>straight sinus occlusion†</td>
<td>moderate</td>
<td>yes/improved</td>
<td>embol/stable</td>
<td>small residual nidus (&lt;1 cm)</td>
<td>14</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>2, M</td>
<td>macrocrania</td>
<td>cerebellar vermis, rt midbrain</td>
<td>3.5</td>
<td>cerebellar veins w/ severe PF congestion</td>
<td>mild</td>
<td>no</td>
<td>NA</td>
<td>none</td>
<td>unchanged nidus (3.5 cm)</td>
<td>11</td>
<td></td>
</tr>
</tbody>
</table>

* Embol = embolization; FU = follow-up; HA = headache; HC = hydrocephalus; NA = not applicable; PF = posterior fossa; pst = posterior; RS = radiosurgery; VeP = venous pouch; VG = vein of Galen.
† In this patient the hydrocephalus was presumably due to venous congestion; the presumed reasons for the hydrodynamic disorder are noted.
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TABLE 2: Literature review of previously reported cases of unruptured pial AVMs in patients with hydrocephalus*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age (yrs), Sex</th>
<th>Location of AVM</th>
<th>DV</th>
<th>Level of Obstruction</th>
<th>VP Shunt</th>
<th>Tx of AVM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lobato et al., 1980</td>
<td>42, F</td>
<td>lt occipital</td>
<td>VG</td>
<td>aqueduct (DV)</td>
<td>yes</td>
<td>none</td>
</tr>
<tr>
<td>Pribil et al., 1983</td>
<td>20, M</td>
<td>rt frontoparietal</td>
<td>rt thalamostriate</td>
<td>foramen of Monro</td>
<td>no op for AVM led to improved HC</td>
<td></td>
</tr>
<tr>
<td>U &amp; Kerber, 1983</td>
<td>31, F</td>
<td>rt thalamus</td>
<td>VG</td>
<td>aqueduct (DV)</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Liu et al., 2003</td>
<td>49, M</td>
<td>lt lower thalamus &amp; midbrain</td>
<td>perimesencephalic</td>
<td>aqueduct (AVM, VeP)</td>
<td>no embol of AVM led to improved HC</td>
<td></td>
</tr>
<tr>
<td>Mindea et al., 2007</td>
<td>55, M</td>
<td>lt parietooccipital</td>
<td>varices, VG</td>
<td>aqueduct (DV, VeP)</td>
<td>yes</td>
<td>RS</td>
</tr>
</tbody>
</table>

* DV = draining vein; NR = not reported.

A less common cause in unruptured AVMs is overproduction of CSF, which has been reported in choroidal AVMs. Obstruction of the ventricles by a drainage vein or the AVM itself has been described previously in only 5 case reports (summarized in Table 2), whereas distant venous outflow obstruction leading to venous congestion, a still hypothetical cause of hydrocephalus in brain AVMs, has not been described for adult pial AVMs.

Obstruction of the ventricles may be caused by the AVM nidus itself, or most commonly by its draining veins. In the latter case, the obstruction can occur at the level of the foramen of Monro or at the aqueduct. Therefore, only AVMs draining into the deep venous system are able to cause ventricular obstruction due to their proximity to the CSF pathways. The veins responsible for this obstruction may be the enlarged thalamostriate-ICV complex or the vein of Galen. Supratentorial AVMs causing this symptom will demonstrate a dominant venous outlet into the deep venous system, whereas infratentorial AVMs will be associated with a dilated vein of Galen. A dilated vein as the cause of obstructive hydrocephalus has been observed in rare cases of complex and large develop-

FIG. 1. Case 1. This 16-year-old girl presented with headaches that had been slowly progressive for 7 months. Plain axial CT scans obtained before (A and B) and after (C and D) VP shunt insertion revealed severe hydrocephalus with periventricular CSF resorption, which remained unchanged after VP shunt placement. Abnormal vessels were noted in the right thalamic region, with an enlarged right lateral perimesencephalic vein draining into a dilated vein of Galen that caused obstruction of the aqueduct due to pressure on the tectal plate (left vertebral artery angiogram, anteroposterior view [E] showing the AVM nidus, which is supplied by thalamoperforating and right posterior choroidal arteries). Embolization of the posterior choroidal feeding vessels (including the venous pouch—seen disappearing on the postembolization angiogram [F]) was performed to reduce the shunt and the size of the AVM, to make the AVM suitable for radiosurgery. Following embolization the patient had clinical improvement of her headaches at follow-up. She is scheduled for radiosurgery.
mental venous anomalies, with a venous collecting vein being located in critical areas as described earlier. Mechanical obstruction by the AVM nidus itself, on the other hand, has only been seen at the infratentorial level for those AVMs surrounding the aqueduct at the level of the midbrain.

In the present collection of cases, we have added 2 patients in whom hydrocephalus was present despite apparent lack of mechanical outflow restrictions. In both cases, though, venous outflow obstruction was observed. Venous outflow restriction resulting in water retention and a subsequent hydrodynamic disequilibrium is typically seen in neonates and infants with vein of Galen AVMs or high-flow pial shunts. In adults, pial high-flow AVMs with venous outflow obstruction have been reported as a possible cause of intracranial hypertension; however, in the reported cases hydrocephalus was not present. In both of our cases, venous outflow restriction may have led to congestion of pial veins of the posterior fossa. Because CSF reabsorption is partly due to a pressure gradient from the ventricles into the pial veins, we hypothesize that the changed venous pressure in the presence of venous outflow restriction and additional arterialization of the veins may have led to a concomitant CSF retention in the ventricles. There have been reports of some patients in whom dural arteriovenous fistulas located at the vein of Galen and confluence of sinuses caused increased pressure in the veins of the posterior fossa, and who presented with the same clinical findings.

Management of the Hydrocephalus: VP Shunt or no VP Shunt?

In our small series we found the results of VP shunt placement to be rather disappointing. There was a high rate of malfunctioning devices due to shunt obstruction, and a high rate of symptoms caused by shunt overdrainage (SDH, hygroma, and slit ventricles), although the opening pressure setting of the programmable valve was...
initially set at a value of 160 mm H2O. In 2 patients, the shunt had no effect on the hydrocephalus (on both imaging findings and clinical symptoms). This unusually high number of complications and shunt failures raises the question whether it may be related to the particularities of the described syndrome.

It is known that patients with vein of Galen AVMs who present with dilated ventricles and who are treated by shunt insertion often develop SDHs, hygromas, or a dilation of the draining vein.1 A possible explanation for this effect is that the pathological mechanism of the hydrocephalus in vein of Galen AVMs is due to an altered hydrodynamic equilibrium, with CSF reabsorption via a pressure gradient from the intraventricular spaces to the medullary veins. Once the pressure in the medullary veins is increased (due to the arteriovenous shunt or additional venous outflow restrictions), less CSF is reabsorbed and the ventricles dilate until a new equilibrium

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**Fig. 3.** Case 4. This 39-year-old woman presented with headaches that had been gradually progressive for 4 months, with new onset of seizures since 3 months before admission. On a T2-weighted MR image (A), abnormal vessels causing obstruction of the right foramen of Monro with unilateral ventricular dilation were demonstrated. An internal carotid artery angiogram (B) revealed an AVM at the right frontoparietal region with dominant deep venous drainage toward the right thalamostriate vein, with a venous pouch at the junction to the ICV. After transarterial glue embolization and partial obliteration of the shunt along with diminution of the AVM size, clinical and imaging improvement of the hydrocephalus was noted, with the patient being free of headaches. The CT scans obtained after embolization (C and D) showed resolution of the hydrocephalus without placement of a VP shunt. The patient underwent further radiosurgery for her residual AVM.
between the intraventricular pressure and the pressure of the medullary veins is found. A VP shunt does not alter the pressure in the medullary veins, but instead just decreases the pressure in the ventricles. These may collapse, resulting in the described hygromas or SDHs; likewise, the draining vein may enlarge. One may argue that the same mechanism may be present in some of the patients presented in this study; an enlargement of the draining vein would lead to an increased pressure on the affected structure, and a lack of effect on the hydrocephalus and the alteration of the hydrodynamic equilibrium would lead to SDHs or hygromas. In this regard, a case report of a dural AVM of the confluence of sinuses in a patient with obstructive hydrocephalus who developed Parinaud syndrome due to upward herniation (probably caused by overdrainage) after VP shunt placement seems noteworthy and along the same line of reasoning.  

Still it has to be pointed out that in the emergency setting, and with acute symptoms of hydrocephalus, the shunt procedure may be the fastest treatment option and therefore must be performed. There are previous reports of successful management of hydrocephalus in an acute setting in patients with neurological deficits by using shunt placement procedures. However, in patients with mild to moderate hydrocephalus without neurological deficits, the aim of treatment (irrespective of the modality chosen) should be reduction of the shunt, with subsequent reduction of the size of the vein causing the obstruction.

Fig. 4. Case 7. This 55-year-old man presented with headaches that had been progressive for 3 months, and sudden worsening of the symptoms, along with vomiting. A contrast-enhanced CT scan (A) showed abnormal vessels at the left cerebellum, with moderate hydrocephalus and generalized brain swelling. Prior to further diagnostic workup, a VP shunt was placed at another institution. After referral to our institution 2 weeks later, the hydrocephalus was still present. Left vertebral artery angiograms in anteroposterior view (B) with a late venous phase in lateral view (C) confirmed the diagnosis of an AVM at the left cerebellum supplied by branches of the left anterior and posterior inferior cerebellar arteries, and draining into the cerebellar veins, with marked stagnation and posterior fossa venous congestion. Nonopacification of the straight sinus is noted, with filling of collateral pathways into the basal vein of Rosenthal. After 2 sessions of transarterial glue embolization that decreased the AVM size by 80%, the patient’s symptoms improved, and he is scheduled for further treatment of the remainder of the AVM. An MR image (D) obtained after the last embolization confirmed reduction of the hydrocephalus, and angiograms (E and F) confirmed the reduction in venous congestion of the posterior fossa.
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In 2 reported cases, the hydrocephalus improved after treatment of the AVM alone (surgical treatment in one and endovascular treatment in the other), as was the case in several patients in our series.

Conclusions

Hydrocephalus is an unusual presentation of unruptured AVMs. Mechanical obstruction by the draining vein or the AVM nidus when located in a strategic position is the most common cause. A rare cause may be hydrodynamic disorders caused by the brain AVM by increasing the pressure within the posterior fossa veins and the deep venous system. Management of the hydrocephalus should be aimed toward treatment of the AVM itself, because only partial improvement is observed following shunt placement procedures. Although VP shunts are necessary in the acute setting, the high rate of associated complications may favor a careful and conservative selection for this treatment among patients who present with this rare symptom.

References


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