Communicating hydrocephalus caused by an unruptured perimedullary arteriovenous fistula in the lumbar region of an infant

Case report


1Faculty of Medicine and Dentistry; 2Division of Neurosurgery, Department of Surgery, and Clinical Department of Neurosciences; and 3Department of Radiology and Diagnostic Imaging, University of Alberta, Edmonton, Alberta, Canada

Spinal arteriovenous malformations are rare in children, although perimedullary arteriovenous fistulas (PMAVs) may account for up to 24% of spinal arteriovenous malformations in this age group. Reported presentations of PMAVs have included progressive or acute myelopathic symptoms, pain, hematomyelia, and subarachnoid hemorrhage. No known reports of an unruptured PMAVF causing communicating hydrocephalus have been previously published.

A 17-month-old girl presented to the authors’ clinic with a 6-month history of back and leg pain, gait regression, constipation, and marked lumbar hyperlordosis due to a PMAVF. A brain MRI study also demonstrated advanced hydrocephalus. The patient underwent embolization with Onyx of 2 feeding arteries from the right L-1 and 1 feeding artery from the left L-1 lumbar arteries. Postembolization follow-up imaging demonstrated a reduction in size of the L-1 pedicles and no residual supply of the fistula. Three-year clinical follow-up showed normal bowel and bladder function with significant improvements in the patient’s back pain, gait, and hyperlordosis. The patient’s ventricular enlargement improved without direct management of her hydrocephalus.

To the authors’ knowledge, this is the first reported case of communicating hydrocephalus caused by an unruptured PMAVF. The authors postulate that the origin of hydrocephalus was either central venous hypertension caused by the high-flow fistula or a change in fluid dynamics reducing CSF resorption through arachnoid granulations in the lumbar region of the spinal cord. The exact role that spinal arachnoid granulations play in CSF resorption is not currently known. Regardless of pathogenesis, initial treatment should focus on management of the fistula with additional hydrocephalus management only when necessary.

(http://thejns.org/doi/abs/10.3171/2012.11.PEDS12244)

Key Words • perimedullary arteriovenous malformation • embolization • perimedullary arteriovenous fistula • hydrocephalus • spine

Intradural extramedullary AVFs, also known as PMAVs, represent a persisting anastomotic connection between intradural spinal arteries and the coronal venous plexus. The overall incidence is unknown, but this type of spinal AVM may account for between 14% and 34% of all spinal AVMs4,16,20,22 and 24% in the pediatric population.9 Presenting signs and symptoms have included acute or chronic progressive myelopathy, sensory abnormalities, bowel and bladder sphincter disturbances, and pain.5,8,18,20,22–24 Hematomyelia and subarachnoid hemorrhages have also been reported.18,20 Single cases of unilateral underdevelopment of the extremities9 and grand mal seizures24 have been described. No known reports of unruptured PMAVs causing communicating hydrocephalus have been previously published.

Case Report

Presentation and Examination. This 17-month-old girl presented to the clinic with a 6-month history of back and leg pain, gait regression, severe constipation, and marked hyperlordosis of the lumbar spine. Prior to deterioration of her condition, the patient’s developmental milestones had been normal. She then regressed from
Communicating hydrocephalus caused by a lumbar perimedullary AVF

normal activity and ambulation, gradually refusing to lie on her back, sit, stand, or walk, mostly because of pain. Her hyperlordosis was fixed, and images obtained of her lower lumbar region demonstrated a 90° curvature.

A spinal MRI study showed a large vascular malformation within the spinal canal, extending from T-10 to the sacrum associated with a large varix at the L1–2 level (Fig. 1A and B). The brain MRI study demonstrated communicating hydrocephalus with transependymal flow (Fig. 2A–C). Subsequent angiography identified a PMAVF fed by bilateral L-1, right L-2, and left L-3 arterial pedicles. A markedly enlarged artery of Adamkiewicz arose from the right L-1 radicular artery with several distal branches supplying the fistula (Fig. 3A and B).

**Treatment.** The patient underwent liquid embolization with Onyx-18 (ev3, Inc.) of 2 arterial pedicles from the right L-1 artery and 1 from the left L-1 artery. After the procedure, no filling of the fistula was demonstrated. A comparative spinal MRI study performed postembolization showed a marked decrease in the size of the venous varix (Fig. 1C and D). The follow-up angiograms showed no residual arteriovenous shunting (Fig. 3C and D).

**Posttreatment Course.** At the 3-year clinical follow-up visit, the patient demonstrated normal bowel and bladder function as well as significant improvements in her gait and hyperlordosis. Objectively, she had normal motor, sensory, and gait examination findings, but she demonstrated persistently brisk lower-extremity reflexes. A brain MRI study showed an improvement of her hydrocephalus with a decrease in ventricular size and complete resolution of transependymal flow (Fig. 2D–F).

**Discussion**

Perimedullary AVFs were first described by Djindjian et al. in 1977 and were officially recognized by Heros et al. as a new category of spinal AVM in 1986. Previous studies reported a relationship between AVMs and hydrocephalus but in a different manner from that presented here. The authors of these studies reported AVMs causing hydrocephalus through several mechanisms, including choroidal AVMs causing increased production and propulsion of CSF, unruptured pial AVMs of the brain causing obstruction, hemorrhage of spinal and brain AVMs causing obstruction, and brain AVMs causing venous congestion resulting in normal-pressure hydrocephalus. None of these mechanisms, however, fully explain how an unruptured malformation in the lumbar spine may induce a communicating form of hydrocephalus.

Discussion on how PMAVFs may be responsible for their reported complications have included 4 potential mechanisms, including mass effect from a venous varicosity, repeated hemorrhage, vascular steal phenomenon, and venous hypertension. Barrow et al. used intraoperative measurements of venous pressure to show that venous hypertension was a potential explanation of the clinical manifestations. Considering the described relationships between AVMs and hydrocephalus, we theorize that this unruptured PMAVF may have caused a communicating form of hydrocephalus by one of the following 2 potential mechanisms: central venous hypertension affecting the hydrostatic pressure of the transgranular CSF flow into the dural venous sinuses, or a change in fluid dynamics involving arachnoid granulations in the lumbosacral region preventing resorption of CSF into the lower spine.

Several authors have reported cases of intracranial dural AVFs presenting with cardiac failure. The presumed cause of associated hydrocephalus in these cases was a central venous hypertension due to poor venous drainage. Theoretically, a large, high-flow PMAVF could cause sufficient central venous hypertension for hy-
drocephalus to result, even if the fistula was found extra-
cranially. The proposed etiology would be a subsequent
decline in the hydrostatic pressure differential between
the subarachnoid space and dural venous sinuses, reducing
the CSF flow across the arachnoid granulations. This could
account for the communicating form of hydrocephalus that
was apparent in our patient.

Alternatively, we considered a change in fluid dynam-
ics involving spinal arachnoid villi as a potential cause
of this communicating hydrocephalus. Spinal arachnoid
villi have been known to exist in animal models,\textsuperscript{11} and
a recent fresh cadaver dissection study has substantiated
their ability to convey CSF to regional veins in humans.\textsuperscript{25}
The extent to which these granulations play a role in CSF
resorption is still not well defined. Tubbs et al.\textsuperscript{25} showed
a significantly greater proportion of arachnoid villi con-
centrated in the lumbar region, the region of our patient’s
venous varix. If spinal arachnoid villi do in fact play a
greater role in CSF resorption than previously thought,
this stands as a potential explanation in our case.

Regardless of the pathophysiology, the literature sug-
ests that management of hydrocephalus caused by AVMs
should focus primarily on the management of the malfor-
mation with CSF shunting considered secondarily.\textsuperscript{10} Geib-
prasert et al.\textsuperscript{10} found that CSF shunting in unruptured brain
AVMs only resulted in partial improvement, although it
was associated with a high complication rate. However,
Kehler\textsuperscript{14} suggested that the high rate of complications seen
in his study could have been reduced had he performed
an endoscopic third ventriculostomy in cases of aqued-
tal compression or had he included gravitational devices

\begin{figure}
\centering
\includegraphics[width=\textwidth]{fig2.png}
\caption{Axial (A, B, D, and E) and coronal (C and F) T2-weighted images of the brain obtained before (A–C) and after (D–F) treatment. The pretreatment images demonstrate ventricular enlargement with mild transependymal CSF flow, which improves on the posttreatment images.}
\end{figure}

\begin{figure}
\centering
\includegraphics[width=\textwidth]{fig3.png}
\caption{Anteroposterior (A) and lateral (B) angiograms of a common L-1 arterial trunk injection demonstrating prominent arterial feeders supplying the artery of Adamkiewicz filling the PMAVF and large venous varix prior to embolization. The anteroposterior (C) and lateral (D) angiograms obtained 3 months after embolization angiographic images (C and D) demonstrate no residual arteriovenous shunting.}
\end{figure}
Communicating hydrocephalus caused by a lumbar perimedullary AVF

to prevent overdrainage of CSF in his shunt-treated cases. Given our patient’s myelopathic presentation and considering our hypothesis for the relationship of the hydrocephalus to the PMAVF, our focus was initially on the management of the fistula rather than the dilated ventricles. Follow-up at 1 and 3 years after surgery has shown no need for additional management of the hydrocephalus.

Conclusions

This is the first known report of an unruptured PMAVF causing communicating hydrocephalus. Two proposals for the pathogenesis of this communicating hydrocephalus have been presented: central venous hypertension decreasing the hydrostatic pressure at the arachnoid granulations, or a change in fluid dynamics in the lumbar region preventing resorption of CSF by spinal arachnoid granulations. The full extent that spinal arachnoid granulations play in CSF resorption is not currently known. Regardless of pathogenesis, treatment of this condition should focus primarily on management of the fistula with additional hydrocephalus management only when necessary.

Disclosure

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 to the study and manuscript preparation include the following. Conception and design: Pugh, Wilson, Aronyk. Acquisition of data: Wilson. Drafting the article: Pugh, Wilson. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Pugh. Administrative/technical/material support: Yeo, Chow.

References


Manuscript submitted May 4, 2012. Accepted November 12, 2012. Please include this information when citing this paper: published online December 14, 2012; DOI: 10.3171/2012.11.PEDS12244. Address correspondence to: Jeffrey A. Pugh, M.D., M.Sc., F.R.C.S.C., 2D1.02 WMC, 8440-112 Street, Edmonton, Alberta, Canada T6G 2B7. email: jppugh@ualberta.ca.