Large vascular malformation in a child presenting with vascular steal phenomenon managed with pial synangiosis

Case report

MICHAEL J. ELLIS, M.D.,1 DEREK ARMSTRONG, F.R.C.P.C.,2 AND PETER B. DIRKS, M.D., PH.D., F.R.C.S.C.1

Divisions of 1Neurosurgery and 2Neuroradiology, Hospital for Sick Children, University of Toronto, Ontario, Canada

The management of large and giant arteriovenous malformations (AVMs) in patients presenting with nonhemorrhagic neurological deficits secondary to vascular steal phenomenon is challenging and controversial. In many cases, large AVMs cannot be completely excised or cured, leaving patients with residual or partially treated AVMs, the natural history of which is unknown. Additionally, large, diffuse vascular malformations with multiple, small feeders, slow flow, or so-called cerebral proliferative angiopathy represent a related but distinct clinical and angiographic entity that may require a different therapeutic approach than traditional brain AVMs. The current management of children with other conditions of chronic cerebral hypoperfusion, such as moyamoya disease, involves consideration of surgical revascularization to enhance blood flow to the compromised hemisphere. Here, the authors present the case of a young child with a large thalamic vascular malformation who presented with clinical and radiological features of vascular steal and ischemia. In an effort to augment flow to the hypoperfused brain and protect against future ischemia, the authors treated the child with unilateral pial synangiosis. At 12 months, postoperative angiography demonstrated robust neovascularization, and the child has not sustained any further ischemic events. The authors discuss concept of vascular malformation–related hypoperfusion and the utility of indirect revascularization for inoperable vascular malformations presenting with ischemic symptoms. (DOI: 10.3171/2010.10.PEDS10388)

KEY WORDS • arteriovenous malformation • pial synangiosis • cerebral proliferative angiopathy • revascularization • vascular steal • children

Abbreviations used in this paper: AVM = arteriovenous malformation; CPA = cerebral proliferative angiopathy; ECA = external carotid artery; ICA = internal carotid artery; MCA = middle cerebral artery; MMA = middle meningeal artery.

The proportion of patients with cerebral AVMs presenting with neurological deficits unrelated to hemorrhage varies widely in reported series.3,11,13,15,26,39 Many reports have attributed these clinical findings to the concept of “vascular steal,” which appears to be mediated by several factors including the size, angioarchitecture, and flow characteristics of the AVM.25 Although the presence of high-flow AVMs may lead to local disturbances in cerebral hemodynamics, the pathophysiology and existence of vascular steal remain in question.25,30 Despite recent advances in our therapeutic armamentarium and encouraging results from experienced centers,3,11,26,39 the management of patients with large AVMs and vascular steal remains challenging and controversial. Patients with large, diffuse AVMs or related CPA are particularly difficult to treat and require individually tailored therapeutic strategies.4,21 In many cases, large vascular malformations cannot be cured, and therapeutic efforts are aimed at modifying specific morphological or flow-related features of the lesion to alleviate progressive neurological symptoms. Unfortunately, partial treatment of AVMs has yet to be shown to alter the natural history of these lesions and may result in poorer outcomes.29,46 In the case of CPA, selected patients may benefit from targeted embolization; however, in some cases, a procedure to enhance blood supply to the hypoperfused cortex may be indicated.21

In children, chronic conditions of impaired cerebral perfusion are rare, the most common of which is moyamoya disease. Children who present with recurrent ischemic symptoms and evidence of impaired cerebrovascular reserve capacity are possible candidates for surgical revascularization in an effort to augment blood flow to the
hypoperfused brain. Several direct and indirect methods have been employed in children including pial synangiosis, which involves the transposition of a branch of the superficial temporal artery onto the brain to promote the gradual formation of supplementary collaterals from the extracranial circulation. In carefully selected patients, pial synangiosis provides safe and durable protection against future ischemic events.

Here, we present the case of a child with a large vascular malformation who presented with clinical and angiographic features of progressive cerebral ischemia indicative of vascular steal. Although the lesion was large, clearly associated with early venous filling, and could be considered to be a “diffuse” or “proliferative angiopathy” type of AVM, we hesitate to refer to as a true or conventional AVM and have thus called it a vascular malformation for the purpose of this report. In an effort to augment flow to the hypoperfused brain and protect against future ischemia, we elected to treat the child with unilateral pial synangiosis. This report lends further support to the concept of vascular steal and offers an alternative treatment for inoperable vascular malformations presenting with nonhemorrhagic neurological deficits or hemispheric hypoperfusion.

Case Report

First Presentation and Examination. This 2-year-old previously healthy girl was referred to the orthopedic surgery service at our institution after falling on her left arm, after which she was noticed to have left upper-extremity weakness. She was seen at the emergency department where she was found to have left-sided weakness involving the upper and lower extremities and a left facial droop. Contrast-enhanced CT scanning of the brain demonstrated a large, densely enhancing mass occupying the entirety of the right thalamus and basal ganglia, with extension into the right temporal and frontal lobe as well as the left thalamus (Fig. 1). Magnetic resonance imaging confirmed the presence of a diffusely extensive vascular lesion associated with extensive T2 signal change in the right frontal, temporal, and parietal regions and significant associated encephalomalacia and atrophy (Fig. 2). No fluid restriction was noted on diffusion-weighted imaging. Catheter-based angiography revealed a 4.5-cm vascular malformation with a complex arterial supply involving the bilateral internal carotid, anterior cerebral, middle cerebral, and posterior cerebral arteries, the right MMA, and the left occipital artery. Venous drainage was early via both normal-caliber veins of Labbé to the transverse sinuses, into the superior sagittal sinus, and into the deep venous system. No dominant high-flow fistulous components were demonstrated. There was significant paucity of opacified distal MCA vessels supplying the right frontal, temporal, and parietal regions on the right internal carotid artery injection consistent with vascular steal. The right ECA injection demonstrated mild parasagittal filling from MMA branches, suggesting a minor extracranial contribution to perfusion of the right medial hemisphere (Fig. 3).

Second Presentation and Examination. The child was seen in clinic weeks later. Her hemiplegia had improved markedly. Given the child’s improved clinical status as well as the vascular malformation’s size and location, diffuse morphology, and varied arterial supply, we believed the lesion was inappropriate for embolization, stereotactic radiosurgery, or resection and elected to follow the patient conservatively. The child was well for 2 years, at which time she presented with dense left-sided hemiplegia. Magnetic resonance imaging demonstrated extensive diffusion restriction involving the right frontal, temporal, and parietal lobes consistent with an acute MCA-territory infarction. The acute infarct was superimposed on a right hemisphere that had undergone significant atrophy. After 3 months of rehabilitation the child exhibited signs of a remarkable recovery, was walking independently with a hemiparetic gait, and her left upper-extremity motor function had returned to her pre-stroke baseline.

Operation. Working on the suspicion that the right cerebral hemisphere was hemodynamically compromised as a consequence of vascular steal from the large vascular malformation and therefore at risk for a future ischemic event, coupled with a good clinical recovery, we elected to perform an indirect revascularization procedure in an effort to augment flow to the compromised hemisphere. The patient underwent a right-sided pial synangiosis, encephalomyosynangiosis, and dural inversion. There were no complications.
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Postoperative Course. A postoperative angiogram obtained 8 months later demonstrated stable configuration of the vascular malformation. Superselective injection of the right ICA and middle meningeal arteries demonstrated enlargement of the superficial temporal artery associated with robust revascularization of the right temporal and inferior frontal and parietal lobe regions with diffuse neovascularization similar to the appearance of the vascular malformation vessels (Fig. 4A–D). At 12 months after revascularization, the patient had sustained no further transient neurological deficits and her left-sided weakness had improved.

Discussion

Focal or progressive neurological deficits unrelated to hemorrhage make up a varying proportion of presenting symptoms in patients with AVMs. Among studies limited to patients with large or giant vascular malformations, the proportion of patients with nonhemorrhagic neurological deficits approaches 50%. Although the exact mechanism underlying this mode of presentation remains unknown, many have attributed it to the concept of vascular steal.

The concept of vascular steal was first described as an angiographic phenomenon characterized by the failure of distal vessels supplying normal brain adjacent to AVMs to opacify due to shunting of blood into the AVM. In this setting, blood is diverted into the low resistance network of the AVM, creating a state of relative hypoperfusion in the surrounding brain that may lead to tissue ischemia. In addition, high pressure in the AVM may also promote venous hypertension leading to impaired venous drainage of the surrounding brain and reduced cerebral perfusion pressure. The resultant ischemia may give rise to clinical sequelae such as transient or progressive focal neurological deficits, seizures, or cognitive deterioration. The angiographic features associated with vascular steal in patients with AVMs include angiomatous change, size, and peripheral venous drainage. In extreme conditions, chronic shunting may give rise to dystrophic intracranial calcifications, a hallmark of chronic hemodynamic disturbance that is also observed in other arteriovenous shunting conditions such as vein of Galen malformations. Despite the finding of decreased cerebral blood flow measurements and imaging findings, the exact mechanism leading to these clinical manifestations remains unclear.

Fig. 2. Coronal T2-weighted MR image revealing an extensive bilateral thalamic vascular lesion with extensive T2 signal change and atrophy of the right frontal, temporal, and parietal lobes.

Fig. 3. Angiograms. A: Anteroposterior view of a right ICA angiogram. B: Lateral view of right ICA angiogram. C: Anteroposterior view of right ECA angiogram. D: Lateral view of right ECA angiogram. Six-vessel catheter angiography revealed a 4.5-cm vascular malformation with an arterial supply arising from bilateral internal carotid, anterior cerebral, middle cerebral, and posterior cerebral arteries, the right MMA, and the left occipital artery (left ICA, left vertebral, and left ECA injections not shown). Significant paucity of opacified distal MCA vessels supplying the right frontal, temporal, and parietal regions on the right ICA angiogram is identified consistent with vascular steal. Minimal transdural supply to the right medial hemisphere is observed from the extracranial system.
paired cerebrovascular reserve capacity\textsuperscript{43} in cortical areas surrounding AVMs, the underlying pathophysiology of vascular steal continues to be debated. While some authors have pointed to impairment in the autoregulatory capacity of arteries supplying adjacent cortex,\textsuperscript{40} others have documented preserved and adaptive autoregulatory responsiveness in these regions.\textsuperscript{17,48,49}

A specific subgroup of patients, who may present with nonhemorrhagic neurological deficits that are important to identify from a clinical and therapeutic perspective, are those with large, diffuse AVMs or CPA. Initially Chin et al.\textsuperscript{4} recognized that a small population of patients with AVMs harbored large, diffuse arteriovenous lesions characterized angiographically by multiple, small arterial feeders, a diffuse “puddling” appearance, and deep venous drainage. They reported on 12 patients, with a mean age at diagnosis of 18.1 years, who presented with hemorrhage (8 patients), seizures (2), headache (1), and nonhemorrhagic neurological deficits (1). Eleven of these patients underwent craniotomy, whereas 1 underwent radiotherapy. Two patients were treated with preoperative embolization. Complete AVM removal was documented in 8 patients, all of whom had excellent clinical outcomes. Pathological examination of surgical specimens demonstrated a striking amount of intervening neural tissue interspersed between abnormal vessels typical of normal AVMs. The authors concluded that these lesions represent an important and unique subtype of AVMs that require carefully tailored management strategies due to the presence of normal and potentially functional neural tissue within these lesions.

More recently, Lasjaunias et al.\textsuperscript{21} described their experience with 49 patients presenting with large, diffuse vascular malformations. The mean age at symptom onset of this population was 22 years; 45% presented with seizures, 41% with severe, often disabling headaches, 12% with hemorrhage, 16% with nonhemorrhagic neurological deficits, and 1 patient presented with hydrocephalus. These cases were characterized angiographically by diffuse, often large, vascular malformations with an absence of dominant arterial feeders, frequent transdural supply, and a classic puddling of contrast material persisting into the venous phase of the angiogram. Although increased arteriovenous transit was described, no high-flow fistulous components characteristic of traditional AVMs were identified. Selected patients underwent perfusion-weighted MR imaging, which demonstrated increased cerebral blood volume and flow within the CPA nidus and increased time to peak and decreased blood volumes in surrounding cortical and subcortical areas suggestive of CPA-related cerebral hypoperfusion. The authors provided the novel term “cerebral proliferative angiopathy” to define this unique entity that appears to differ significantly from normal brain AVMs in terms of epidemiology, clinical presentation, natural history, and management considerations. They concluded that treatment be restricted to those patients presenting with hemorrhage, uncontrolled seizures, and disabling headaches and cautioned against the use of nontargeted embolization, which could place the functional intervening neural tissue at risk for embolization. Due to the ischemic nature of this disease, they suggested that selected patients may benefit from therapies that enhance blood flow to the hypoperfused cortex. In this series, calvarial bur holes were made in 2 patients and clinical improvement followed.

Among other conditions that can present with transient or progressive neurological deficits as a result of impaired cerebral perfusion is moyamoya disease. Moyamoya disease is a idiopathic, noninflammatory vasculopathy characterized by progressive stenosis or occlusion of the supraclinoid internal cerebral arteries with resultant arborization of abnormal leptomeningeal collaterals.\textsuperscript{41,42} Children with moyamoya disease typically present with recurrent transient ischemic attacks or ischemic stroke, whereas adults often present with hemorrhagic stroke. Functional imaging studies have documented impaired patterns of ce-
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rebral blood flow and cerebrovascular reactivity in moy-

amoya disease. Patients who present with hem-

orrhage or recurrent ischemic events in the presence of

impaired cerebrovascular reactivity are often considered

for surgical treatment. Direct and indirect revasculariza-
tion techniques have been described, all with the common

goal of enhancing blood flow to the hypoperfused brain. While

direct revascularization for vascular steal phenomena

could not be offered without a high risk of morbidity and
death. The lesion in our patient may have a lower bleeding

natural history than high-flow brain AVMs. As such, we

elected to perform a unilateral pial synangiosis in an effort
to augment flow to the compromised hemisphere. At the

8-month follow-up, postoperative angiography demonstrat-
ed robust revascularization of the right temporal, inferior

frontal, and parietal lobe regions with diffuse neovascular-
similar to the appearance of the vascular malfor-
mation vessels. Since the revascularization procedure she

has not suffered another clinical ischemic event. Indeed, we
do wonder, in light of her established cerebral atrophy
and the minimal transdural supply to her right hemisphere
from the extracranial circulation, if her acute stroke may
have been prevented had we offered a revascularization
procedure earlier. To our knowledge, this represents the

first reported case of a vascular malformation presenting

with vascular steal treated with pial synangiosis. While it

is conceivable that the robust revascularization in the ex-
tracranial circulation may serve to augment flow to the

patient’s symptomatic hemisphere, functional cerebrovas-
cular reserve studies were not carried out in this case, so

this notion remains only speculative. Surprisingly, the

neovascularization that resulted from the pial synangiosis

was very similar to that of the malformation vasculature

and unlike that which is typically observed following indi-
rect revascularization for moyamoya disease. Although the

sump effect of the large vascular malformation may have

contributed to the acquisition of the abnormal pattern of re-
vascularization, the extracranial circulation was not found
to contribute to early venous filling or filling of the thal-
lamic nidus. Whether this revascularization pattern reflects

an abnormal angiogenic environment within the hypoper-
fused hemisphere or an underlying abnormality affecting

mediators of cerebral vessel formation in this patient is

unknown. Indeed, levels of vascular endothelial growth

factors and other proangiogenic proteins have been shown
to be upregulated in AVM endothelial cells and surround-
ing astroglia. Elevated levels of basic fibroblast growth

factor, a mediator of angiogenesis, have also been detected

in the CSF of patients with moyamoya disease, with higher

levels associated with more robust angiographic outcomes

from pial synangiosis.

Conclusions

Given the challenges of treating large, diffuse AVMs and
CPA presenting with vascular steal and neurological
deficits, this report offers an alternative mode of treatment
that may be used to augment flow to the hypoperfused
cortex without causing abrupt disturbances in cerebral
hemodynamics. In this setting, indirect revascularization
procedures, such as pial synangiosis, appear ideal as neo-
vascularization occurs over time and seems to be governed
by the physiological need and angiogenic milieu of the
hemodynamically compromised hemisphere. Long-term
follow-up is warranted to assess whether this technique
can offer durable protection from further ischemic insults
in this setting.

Disclosure

The authors report no conflict of interest concerning the mate-

Author contributions to the study and manuscript preparation include the following. Conception and design: Dirks, Ellis. Acquisition of data: Ellis, Armstrong. Analysis and interpretation of data: Ellis, Armstrong. Drafting the article: Dirks, Ellis. Critically revising the article: Dirks. Reviewed final version of the manuscript and approved it for submission: all authors. Administrative/technical/material support: Dirks. Study supervision: Dirks.

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Manuscript submitted September 1, 2010. Accepted October 14, 2010. Address correspondence to: Peter B. Dirks, M.D., Ph.D., Hospital for Sick Children, Room 1503, 555 University Avenue, Toronto, Ontario, Canada MSG1X8. email: peter.dirks@sickkids.ca.