Capillary hemangiomas of the central nervous system are composed of a compact arrangement of capillaries lined by plump or flattened endothelial cells without intervening neural tissue, possessing histological features similar to those of lobular capillary hemangioma of the skin and capillary hemangiomas of infancy, except that intracranial capillary hemangiomas tend to be more cellular than their cutaneous counterparts. Intracranial capillary hemangiomas (ICHs) are distinctly different from cavernous malformations, distinguishable by their appearance on imaging, propensity for hemorrhage, and histopathology. MRI of cavernous malformations tends to display an intraparenchymal lesion with little to no enhancement, evidence of chronic prior hemorrhage, and rarely shows signal changes consistent with extensive surrounding edema. This is in contradistinction to ICHs on MRI that are often not intraparenchymal, are brightly enhancing, infrequently show evidence of prior hemorrhage, and can be associated with extensive perilesional edema. Clinically it is unusual for patients with an ICH to present because of hemorrhage, whereas hemorrhage is a common cause of clinical presentation in patients with cavernous malformations. Unlike ICHs as described above, cavernous malformations, histopathologically, have dilated capillaries with thin-walled endothelial cells and often have evidence of prior hemorrhage. ICHs are considered at the benign end of the spectrum of proliferative vascular lesions that include papillary endothelial hyperplasia, hemangioendothelioma, cellular hemangioma, hemangiopericytoma, and angiosarcoma. They are considered more hamartomatous than neoplastic.

ICHs come to clinical attention infrequently. Presentation falls predictably into three categories as with most intracranial masses: symptoms of raised ICP, seizure, or cranial nerve or motor deficit. Only 14 children with ICHs have been reported in the literature (Table 1). Two more pediatric cases of ICH are reported here in conjunction with a review of the literature to put forth a perioperative strategy and highlight the surgical challenges and lessons learned with these exceedingly rare intracranial lesions.

Case Reports

Case 1

A 10-year-old boy presented with a 2- to 3-week history of worsening headache. He was an otherwise completely healthy child. There was no family history of intracranial neoplasms or vascular malformations.

Examination

His cardiac and cutaneous examinations were normal. He had mild papilledema, and no focal neurological defi-
<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Location</th>
<th>MRI Findings</th>
<th>Angiographic Findings</th>
<th>Treatment</th>
<th>Outcome</th>
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<tr>
<td>Abe et al., 2005 &amp; 2004</td>
<td>8, M (1st case); 16, F (2nd case)</td>
<td>Temporal lobe (1st case); multiple ICH (2nd case)</td>
<td>Both w/ signal changes consistent w/ perilesional edema. All lesions markedly enhanced w/ Gd</td>
<td>Both w/ hypervascular masses w/ pial supply</td>
<td>Surgery in both cases</td>
<td>Complete resection (1st case); complete resection &amp; regression of remaining lesions after 5 mos (2nd case)</td>
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<tr>
<td>Brotchi et al.</td>
<td>10, F</td>
<td>Occipital, sagittal</td>
<td>Brightly enhancing, dural based</td>
<td>Hypervascular, ECA supply</td>
<td>Surgery</td>
<td>Small residual tumor</td>
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<td>Daenekindt et al.</td>
<td>0.1, M</td>
<td>Temporal</td>
<td>Brightly enhancing</td>
<td>Hypervascular, meningeal arterial supply</td>
<td>Biopsy, embolization, &amp; surgery</td>
<td>Complete resection</td>
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<td>Frei-Jones et al.</td>
<td>0.01, F</td>
<td>Middle fossa, posterior fossa</td>
<td>Brightly enhancing w/ prior hemorrhage</td>
<td>Not done</td>
<td>Biopsy, thalidomide</td>
<td>&gt;95% reduction in size at 20 mos</td>
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<tr>
<td>Karamchandani et al.</td>
<td>0.25, M</td>
<td>4th ventricle</td>
<td>Brightly enhancing w/ prior hemorrhage</td>
<td>Not done</td>
<td>Surgery</td>
<td>Complete resection</td>
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<tr>
<td>Karikari et al.</td>
<td>0.1, M</td>
<td>Medial temporal lobe, anterior choroidal artery</td>
<td>Hypointense on T2WI. “Vascular-type lesion compatible with aneurysm or arteriovenous malformation” by MRA</td>
<td>Not done</td>
<td>None/autopsy</td>
<td>Death</td>
</tr>
<tr>
<td>Mirza et al.</td>
<td>14, M</td>
<td>Middle &amp; infratemporal fossa seemingly arising from w/in petrous bone</td>
<td>Brightly enhancing &amp; encasing petrous carotid artery</td>
<td>Hypervascular w/ ECA supply</td>
<td>Embolization, subtotal resection, &amp; irradiation</td>
<td>“Tumor control”</td>
</tr>
<tr>
<td>Morace et al.</td>
<td>8, M (1st case); 13, M (2nd case)</td>
<td>Occipital/tentorial, temporal/occipital/tentorial</td>
<td>Brightly enhancing w/ signs of hemorrhage; significant perilesional edema (1st case); brightly enhancing w/ significant perilesional edema &amp; cyst (2nd case)</td>
<td>Not done (1st case); hypervascular w/ ECA supply (2nd case)</td>
<td>Surgery in both cases</td>
<td>Near-complete resection (1st case); small progression of residual (2nd case)</td>
</tr>
<tr>
<td>Phi et al.</td>
<td>15, F</td>
<td>Cavernous sinus</td>
<td>Brightly enhancing</td>
<td>Not done</td>
<td>Biopsy, irradiation</td>
<td>Near-complete resolution</td>
</tr>
<tr>
<td>Simon et al.</td>
<td>0.33, F</td>
<td>Cerebellum</td>
<td>Cystic mass</td>
<td>Not done</td>
<td>Surgery</td>
<td>Complete resection</td>
</tr>
<tr>
<td>Tsao et al.</td>
<td>1.5, M</td>
<td>Frontal/temporal</td>
<td>Brightly enhancing, dural based</td>
<td>Hypervascular w/ middle meningeal supply &amp; early draining vein</td>
<td>Surgery</td>
<td>Complete resection</td>
</tr>
<tr>
<td>Willings et al.</td>
<td>3, M</td>
<td>Frontal/temporal/parietal</td>
<td>CT only</td>
<td>Not done</td>
<td>Surgery</td>
<td>Complete resection</td>
</tr>
</tbody>
</table>

* ECA = external carotid artery; MRA = MR angiography; T2WI = T2-weighted imaging.
cits. His visual fields were normal. An occipital lesion based on the tentorium was diagnosed preoperatively (Fig. 1). The lesion enhanced brightly and was sharply circumscribed. There was significant perilesional edema. Flow voids, draining veins, and signs of prior hemorrhage were absent on the imaging studies.

Operation

He underwent an image-guided occipital craniotomy. Despite preoperative dexamethasone, intraoperative mannitol, mild hypocarbia, head elevation, and no pronounced neck flexion, there was profound occipital lobe swelling. The occipital lobe, however, was able to be safely elevated off the tentorium-based lesion while utilizing the intraoperative microscope. A bright red, obviously vascular mass with numerous arterial feeders and a red draining vein was encountered. Although the MRI findings were not consistent with an arteriovenous malformation, there was the concern of failure to recognize a high-flow vascular lesion preoperatively given the intraoperative appearance. It was decided to halt the case and obtain an angiogram under the same anesthetic. The dura could not be closed because of brain swelling and the bone flap was not fixated but left loose under the scalp to accommodate the occipital lobe swelling. The angiogram with both vertebral, external carotid, and internal carotid artery injections displayed only a faint blush on the vertebral injection, but no early draining vein (Fig. 2). Intravenous dexamethasone was administered between operations. The child was returned to the operating room the next day and the lesion was resected. Numerous feeders from the tentorium were coagulated and the lesion removed en bloc. The occipital swelling at this point was inconsequential and dura and bone flap were closed and fixated, respectively. No blood transfusion was necessary for either operation.

Postoperative Course

The child’s neurological examination was normal. Pathological findings were diagnostic for capillary hemangioablastoma, and this diagnosis was corroborated by pathological review at another academic institution (Fig. 3). Contrast-enhanced cranial MRI 30 months postoperatively showed no evidence of residual or recurrent lesion (Fig. 4).

Case 2

A 14-year-old boy presented with worsening diplopia a few days following a minor bicycle accident. He was a healthy child by history. There was no family history of intracranial neoplasms or vascular malformations.

Examination

The patient’s cardiac and cutaneous examinations were normal. On neurological examination he was alert and interacted appropriately. There was a left abducens nerve paresis, and mild left central facial weakness. A mass based in the region of the sylvian fissure was diagnosed preoperatively (Fig. 5). Like the imaging features in Case 1, the almost 3-cm lesion enhanced brightly and was sharply circumscribed. There was significant perilesional edema. Flow voids, draining veins, and evidence of prior hemorrhage were absent on imaging.

FIG. 1. Case 1. A: Coronal T1-weighted Gd-enhanced MR image showing brightly enhancing well-circumscribed mass based on the tentorium with perilesional signal change consistent with edema. B: Axial FLAIR image demonstrating left occipital signal change consistent with edema.

Operation

The patient underwent an image-guided right frontoparietal craniotomy. Again, despite preoperative dexamethasone, intraoperative mannitol, hypocarbia, neutral neck position, and head elevation, there was profound cerebral swelling. After durotomy, the mass auto-delivered itself through the sylvian fissure as the brain mushroomed up through the dural opening. Intraoperative ultrasound was used to look for an intraparenchymal hematoma, but no satisfactory images could be obtained with the probe positioned atop the mushrooming brain. Once again utilizing the intraoperative microscope, visualization of the lesion showed it to be a bright red, obviously vascular lesion similar to a varix containing oxygenated blood and numerous communications both, venous and arterial, with the sylvian vessels. Any mobilization of the lesion brought forth brisk bleeding from the sylvian fissure. Again, there was concern that there was failure to recognize a high-flow vascular lesion preoperatively, and it was decided to halt the case and obtain an angiogram under the same anesthetic. Additionally, since the brain swelling was so profound, cranial CT was performed to assess for intracranial hemorrhage deep to the lesion. The craniotomy was expanded and the dura widely opened without replacement of the bone flap before closing. The CT scan revealed a hematoma medial and superior to the mass, along with the known brain swelling (Fig. 6). Internal and external carotid artery angiograms displayed no abnormal blush or early draining vein (Fig. 7). The child was immediately returned to the operating room where the lesion was resected en bloc. Numerous vessels spanned between the sylvian branches and the lesion. There was one significant arterial supply coming from a branch of the middle cerebral artery (MCA) that was either avulsed in association with the spontaneous herniation of the lesion upon dural opening or in association with microsurgical manipulation. Given the near auto-delivery of this mass upon simply opening the dura, the former etiology of spontaneous avulsion was suspected. This avulsion from a branch of the MCA was the source of the hematoma depicted on the
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CT scan as the blood that jetted under the medial aspect of the mass and superiorly. There was no stump to coagulate but only a small hole in the MCA branch jetting arterial blood. This was covered with hemostatic agents and slight pressure was added with a cottonoid. After several minutes, there was no bleeding with the release of pressure. Despite preservation of the major sylvian vasculature, complete removal of the lesion, and evacuation of the hematoma, the brain swelling remained profound. The craniotomy was further enlarged, the dura opened more widely, the brain covered with a dural substitute, and the scalp closed without bone replacement. No blood transfusion was necessary.

Postoperative Course

After surgery, the patient had mild transient worsening of his central facial paresis, which has since completely resolved. Pathological features were diagnostic of capillary hemangioma. The diagnosis was corroborated by pathological review at another academic institution (Fig. 8). Cranioectomy was scheduled for approximately 10 weeks after the initial craniotomy, but the cerebral edema was still too profound to place a prosthetic flap. Not until 6 months postoperatively did the cerebral edema clinically and radiologically diminish to allow for cranioplasty. Two years postoperatively he experienced a seizure after staying up all night. Academically he has performed no differently than prior to diagnosis. Immediate postoperative and long-term follow-up imaging showed no residual or recurrent mass but did display the brain swelling and encephalomalacia, respectively (Fig. 9).

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As reinforced by the 2 cases presented here, there is a predilection for more boys to be diagnosed with ICH in the pediatric age group. Including these 2 cases, the boy/girl ratio is 11:5 for pediatric intracranial ICH (Table 1). When ICH presents in adulthood there is a predilection for women to be affected. In the 18 cases reviewed by Phi et al., of both children and adults with ICH, males presented at a mean age of 4.8 years and females at a mean age of 22.5 years. While not all 14 patients reported on in the literature underwent a Gd-enhanced MRI, of those for whom the MRI findings were adequately described in the published reports, 9 had brightly enhancing lesions and 4 had significant perilesional edema. It is possible more than 9 and 4, respectively, had lesions with bright enhancement or significant perilesional edema, as in some reports either the representative images or their description was lacking.

The 2 patients described here had clinical presentations typical of raised intracranial pressure (ICP) due more to the profound edema than mass effect from the ICH itself. The cause of this edema is not clear. From this experience and the literature, ICHs associated with the tentorium or those found within the parenchyma tend to be associated with edema. Those ICHs that seemingly arise from the skull base or have wide dural bases mimicking meningiomas tend not to have associated edema (Table 1). It would be tempting to surmise that the lesion contains a high concentration of capillaries without a normal blood-brain barrier and this nest of leaky capillaries causes the edema. These lesions also may be associated with yet-to-be-defined abnormalities of the normal venous drainage, as seen with the coexistence of developmental venous malformations and cavernous malformations causing regional venous hypertension. This may explain why the second child required a prolonged time for edema resolution. Finally, as shown by Frei-Jones et al., these lesions can be associated with elevated vascular endothelial growth factor levels that may affect the capillary integrity of the blood-brain barrier within the surrounding brain.

Additional causes of raised ICP other than edema in ICH have been reported as well. Brotchi et al. have described the case of a child presenting with elevated ICP secondary to an ICH that caused sagittal sinus obstruction. Other authors have reported children presenting with signs and symptoms of elevated ICP secondary to the shear size of the ICH itself. One infant died of a subarachnoid hemorrhage in association with a skull base ICH fed by the anterior choroidal artery.

While few cases of ICH have been described, the MRI
features at presentation in the 2 cases reported here were consistent with those in many prior reports of adults and children. A brightly enhancing, well-circumscribed lesion on T1-weighted images with Gd in close proximity to major vascular channels was seen in both cases. On T2-weighted or FLAIR images, signal change consistent with extensive perilesional edema was also present in both cases. Two reports have described lesions mimicking convexity meningiomas with broad-based dural attachments without cerebral edema. The case of a fourth ventricular ICH in a 3-month-old child presenting with hypotonia had no associated edema. Most reported cases of ICH in which angiography was performed preoperatively showed a significant blush of contrast filling in the capillary phase. Both pial and external carotid arterial supplies have been described. The angiographic findings seen in the 2 cases reported here, however, may have been distorted by the fact the angiograms were obtained after craniotomy, but before resection, and a degree of devascularization may have occurred. In the present 2 cases angiography showed either a subtle blush in the capillary and venous phases or no detectable contrast filling. Some ICHs have been embolized preoperatively to assist with resection.

While also exceedingly rare, supratentorial hemangioblastomas would be considered in the differential diagnosis. The MRI findings of hemangioblastomas tend not to show the profound perilesional edema seen with ICHs, and vascular flow voids are often displayed. Association with cysts is common with hemangioblastomas, but not with ICHs, though 2 pediatric cases did have ICHs associated with cysts. The angiographic findings of hemangioblastomas tend to be more dramatic, with a very dense tumor blush, dilated feeding arteries, and dilated draining veins. In one child with an ICH, the angiographic findings shared similarities with those of hemangioblastomas. Papillary endothelial hyperplasia (PEH) is another vascular proliferative disorder to be considered in the differential diagnosis and is also exceedingly rare. PEH is thought to be an exuberant form of organizing thrombus that can arise in a preexisting vascular abnormality (intravascular) or hematoma (extravascular). MRI of PEH shows poor to variable enhancement, with evidence of old hemorrhage and variable amounts of mild edema. Histopathologically, numerous vascular channels surround fibrin cores. Evidence of preexisting hemorrhage and the histopathological characteristics of PEH were not seen in the 2 present cases. Angiography in the setting of a possible ICH serves multiple purposes of excluding vascular anomalies such as arteriovenous malformation or aneurysm that may be treated by nonsurgical means, allowing for embolization in selected lesions, and providing the angioarchitecture of the lesion for the surgeon. Having angiographic information preoperatively in these 2 cases would have simplified the surgical management because the surgeon would have known that the lesions were not high-flow vascular lesions despite their intraoperative appearance.

Most reported cases have been treated with complete surgical resection. Because of the vascular nature of these lesions, either open or stereotactic biopsy would be ill advised were the lesion considered to be an ICH preoperatively. Some reports, however, have described partial resection of ICH in adults and children. These partially resected lesions often involved the skull base and cavernous sinus. The residual portion was then successfully treated with fractionated radiation.

Two cases of patients with multiple ICHs have been...
treated successfully with resection of one lesion and medical management of the remaining lesions (steroids and alpha-interferon). The response of cutaneous capillary hemangiomas in infancy to exogenous steroids is a well-recognized treatment to avoid surgery or to shrink lesions before undertaking surgery. Steroid receptor positivity was seen in 3 of 10 lesions involving the brain or spinal cord. Whether steroid receptor presence correlates with a therapeutic response to glucocorticoids is not known. One infant with a giant lesion that could not be safely resected was successfully managed by undergoing a 19-month course of thalidomide, an antiangiogenic agent, which shrank the lesion to a small residual mass. Treatment response correlated with systemic vascular endothelial growth factor levels. Similarly using another antiangiogenic agent, bevacizumab, along with temozolomide, an intracranial cellular hemangioma in an infant was successfully treated. Cellular hemangioma is considered a more aggressive lesion but is related to ICH along the spectrum of vasoproliferative lesions.

As for the 2 cases in the present report, the lessons learned are several. While the preoperative imaging was “typical” for ICHs, these lesions are exceedingly rare and an ICH was not considered preoperatively. Given the vivid enhancement with gadolinium and the lesion’s proximity to major vascular structures, some type of angiographic imaging would have been helpful preoperatively to better understand the vascular nature of the lesion in each case and provide confidence that a high-flow vascular lesion was not present. Formal angiography has been used successfully in reported cases more than MR angiography. Angiography also allows for embolization if indicated. There have been no reported cases in which CT angiography was performed.

Preoperative consideration of ICHs in the differential diagnosis could have allowed for preoperative medical management with steroids or other agents discussed above to possibly reduce the profound cerebral edema associated with these lesions and thus make the operation easier and with less morbidity. The exact medical regimen, however, would have been empirical and not necessarily successful.

Also given the edema present with these relatively small lesions, a wide surgical field should be draped. While neurosurgeons may take pride in removing intracranial lesions through the smallest of craniotomies, with a potential ICH the surgeon should be prepared to undertake a large decompressive craniotomy if intraoperative brain swelling otherwise cannot be controlled.

While ICHs seem not to present because of hemorrhage, intraoperative spontaneous or surgeon-induced bleeding (e.g., from a biopsy) can be an intraoperative complication requiring an array of hemostatic agents and need for volume, blood, and coagulation factor replacement. Hence, the surgical and anesthetic teams should be prepared beyond the normal preparation for a routine pediatric brain tumor case.

Conclusions
ICHs are benign vascular lesions that can be surgically treacherous because of their vascularity and associated brain edema. A brightly enhancing, well-circumscribed mass shown on MRI near the dural sinuses or major vascular structures, especially with significant perilesional edema, should alert the surgeon to the possibility of an ICH. Appropriate imaging studies to exclude high-flow vascular lesions preoperatively are indicated if an ICH is in the differential diagnosis. If the diagnosis is known through biopsy, incomplete resection, or complete resection of one of multiple ICHs, then medical and radiotherapy options seem effective for residual lesions. Preoperative medical regimens to reduce brain swelling should be considered. The surgeon and anesthetic team should be prepared for spontaneous hemorrhage, substantial blood loss, and difficult-to-control brain swelling. Surgical excision is curative.

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References


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