Intracranial arteriovenous malformations are believed to be congenitally acquired lesions. Their association with brain neoplasms is extremely rare, and data are limited to case reports.\textsuperscript{4–11,13,14,21–23} It has been postulated that the hyperangiogenic environment of high-grade tumors induces abnormal arteriovenous connections.\textsuperscript{7} It is not known why only a very few brain neoplasms develop large arteriovenous shunts with vascular niduses mimicking true AVMs. We report such a lesion found within an oligodendroglioma in a patient who had recently undergone brain imaging with unremarkable results, and we describe the treatment of this unique lesion with endovascular embolization followed by excision.

Case Report

History. This 55-year-old woman presented with a left-sided thalamic hemorrhage in October 2002. No vascular imaging was obtained and she recovered without sequelae. In July 2003, she presented with transient left-sided weakness. Brain MR imaging (Fig. 1A) showed old blood products in the left thalamus (arrow) but no acute infarct. Findings on intracranial MR angiography (Fig. 1B) were unremarkable. In November 2005 the patient presented again with acute onset dysarthria, left-sided weakness, and left visual field deficits. Admission MR imaging revealed an enhancing 5-cm right frontoparietal mass with vasogenic edema, midline shift, and significant flow voids (Fig. 1C and D). A diagnostic cerebral angiogram confirmed the presence of high-flow arteriovenous shunting and a vascular nidus with feeding vessels supplied by branches of the right middle cerebral artery (Fig. 2A and B).

Operation. A decision was made to perform a staged embolization followed by craniotomy and either an open biopsy (if the lesion were found by frozen section to be a malignant neoplasm) or excision (if no neoplasm was found on frozen pathological specimens). The arteriovenous lesion was treated with N-butyl cyanoacrylate glue injections, with good angiographically confirmed reduction in the flow and velocity of blood through the nidus.
shunt (Fig. 2C). A repeated angiogram with possible sec-
ond-stage embolization demonstrated no readily acces-
sible arterial target to embolize.

At surgery the brain appeared swollen, with signifi-
cant sulcal effacement. Frozen-section specimens failed
to show neoplasm, and gross-total removal of a presumed
nonneoplastic AVM was pursued. No glial plane was iden-
tified, and active aggressive bleeding was frequently en-
countered that was difficult to control. After several hours
of surgery with resistant bleeding, the brain began to herni-
ate outward, with malignant swelling. A ventriculostomy
was placed to treat obstructive hydrocephalus from intra-
ventricular blood. Ventricular drainage resulted in brain
relaxation, and a gross-total resection was achieved. The
patient was placed in a pentobarbital coma for cerebral
protection and to control the blood pressure.

Postoperative Course. A right carotid angiogram
performed after the resection showed no evidence of re-
sidual vascular shunting (Fig. 2D). Venous thromboembo-
lism prophylaxis consisted of compression stockings and
pneumatic boots. Anticoagulants were not used. Despite
negative findings on lower-extremity duplex studies, the
patient suffered a massive pulmonary embolism and died
on postoperative Day 6. No postoperative MR imaging or
neurological examination after recovery from pentobar-
bital sedation were obtained. No autopsy was performed.

Discussion

Although they are extremely rare, arteriovenous les-
sions with large shunts and a vascular nidus mimicking
AVMs may be associated with intracranial neoplasms.
The term “angioglioma” was first used by Councilman in
1914 to describe cerebellar hemangioblastomas, but has
been used more broadly to describe neoplasms in-
volving vascular malformations. Despite the few reports
of hemangioblastomas and meningiomas associated
with arteriovenous shunts, the great majority of dual-
pathology lesions composed of arteriovenous shunts and
neoplasms have involved tumors of glial origin.
The existing literature describing arteriovenous shunts and gliomas diagnosed simultaneously as a combined lesion consists of a few case reports. These reports suggest an association between such lesions and primary brain neoplasms, specifically oligodendrogliomas and astrocytomas. However, this association is rare and their existence as a distinct entity continues to be debated.

To better understand arteriovenous shunts associated with brain tumors, Lombardi et al. performed a retrospective histological analysis of 1034 AVMs treated at one institution. In this study they did not find any neoplastic element, but described 8 cases (0.1%) of increased oligodendroglial cell proliferation, which was further characterized by 2 nonneoplastic histological patterns. The cellular prominence in 3 cases was thought to be “malformative,” resulting from developmental changes related to the formation of the AVM. The histological appearance of the 5 other cases was termed a “collapse pattern” of tissue causing a relatively hypercellular environment.

Lombardi et al. further reviewed the histopathology of 82 oligodendrogliomas and 104 cerebellar and 51 supratentorial pilocytic astrocytomas. A small subset of these tumors, 4 (5%) of the 82 oligodendrogliomas, 5 (5%) of the 104 cerebellar pilocytic astrocytomas, and 6 (12%) of the 51 supratentorial pilocytic astrocytomas were found to be highly vascular, but none had angiographically confirmed arteriovenous shunting.

In their small case series, Nazek et al. reviewed the neuropathological findings in 3 AVMs that showed oligodendroglial cell proliferation on the margins of the malformation, and concluded that this did not meet sufficient criteria to be considered neoplastic. They suggested that the findings in other reports suggesting dual-pathology AVM and oligodendroglioma might have been inaccurate and overstated. Despite the skepticism, there are reported cases that have documented angiographically confirmed arteriovenous shunts associated with pathologically confirmed neoplasms.

A literature review revealed 13 earlier cases of arteriovenous shunts associated with tumors of glial origin, including 3 oligodendrogliomas, 7 astrocytomas, 1 “high-grade” glioma, 1 glioblastoma multiforme, and 1 pleomorphic xanthoastrocytoma (Table 1). Nine of the cases were anatomically indistinct lesions, and 3 were noncontiguous but involved the same hemisphere, and 1 occurred in the contralateral hemisphere. Only 2 other cases have been reported since the advent of MR imaging. In none of the others was there documentation of de novo formation of simultaneous glioma and large arteriovenous shunting and nidus mimicking an AVM. Arteriovenous shunting was documented by cerebral angiography in 12 of 14 cases. Methods of treatment included preoperative endovascular embolization in our case alone, gross-total or subtotal resection in at least 9 cases, and radiotherapy in at least 4 cases. Few long-term data are reported; however, 3 of the patients died within 14 months of their diagnosis.

Our case is the first in which previously acquired, negative brain MR images were available. The absence of tumor or abnormal vasculature on MR imaging and MR angiography obtained 27 months prior to the patient’s pre-
sentation is intriguing given the widely held belief that arteriovenous shunting and the presence of a vascular nidus as is found in AVMs are congenitally derived. However, some previous studies have challenged this concept.\(^1\,\,\,^7\,\,\,^19\,\,\,^20\)

A case report by Harris et al.\(^7\) documented growth of an arteriovenous lesion in the setting of a high-grade astrocytoma on serial angiograms. They postulated that the tumor created a hyperangiogenic environment that fostered growth of the aberrant vasculature. An alternative theory is that an environmental exposure induced the formation of both lesions.\(^2\)

We found no technical difference between the embolization of this lesion and nonneoplastic AVMs. Endovascular embolization has been used successfully in the adjunctive treatment of other vascular intracranial neoplasms without associated AVMs.\(^17\) Despite technically routine embolization, resection of such lesions appears hazardous, and significant blood loss may be expected given the lack of a glial plane usually associated with AVMs. Coagulation of the vessels was more difficult than it was in AVMs, and perhaps reflects their neoplastic involvement. Given the dangers associated with attempted resection in such lesions, special consideration should be given to open biopsy followed by radiation if the frozen section shows malignant pathological features.

A lesion with large arteriovenous shunting, a vascular nidus, and neoplastic cellular proliferation may be a distinct entity, although this may simply represent the end of a spectrum of vascularity known to be associated with malignant gliomas. Given the incidence of AVMs and neoplasms in general, it is not surprising that on rare occasions an unfortunate individual might by coincidence harbor both lesions. This would, however, more readily explain the situation in which the lesions are separate from one another. The serial neuroimaging in our case and that of Harris et al.\(^7\) provide strong evidence that such arteriovenous lesions were not congenitally derived and that their growth was unlikely to have been coincidental with that of the tumor. This case also provides evidence that large arteriovenous shunting with a vascular nidus within the brain can develop in < 27 months in the proper environment. Although we did not pursue such a study in this case, in the future molecular evaluation of neoplastic arteriovenous lesions might provide genetic information that could be used to study the origin of AVMs and assist with the creation of a true animal model of brain AVM, which currently does not exist.

**Conclusions**

This is the first case report to demonstrate the de novo occurrence of an arteriovenous lesion with large shunts and a vascular nidus within an anaplastic oligodendroglioma in a patient with previously unremarkable imaging findings. Although embolization for these entities may be technically indistinguishable from that of nonneoplastic arteriovenous lesions, surgery is extremely hazardous given the high vascularity, neoplastic infiltration of the vessel walls, and lack of a glial plane normally found in brain AVMs.

### TABLE 1

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>AVM/Tumor Location</th>
<th>Radiographic Study</th>
<th>Treatment</th>
<th>Tumor Histology</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fine et al., 1960</td>
<td>15, M</td>
<td>rt parietal/ventricular</td>
<td>ventriculogram &amp; angiogram</td>
<td>GTR &amp; XRT</td>
<td>oligodendroglioma</td>
<td>NA</td>
</tr>
<tr>
<td>Wecker &amp; Seidel, 1966</td>
<td>25, F</td>
<td>rt parietal</td>
<td>angiogram</td>
<td>NA</td>
<td>astrocytoma</td>
<td>NA</td>
</tr>
<tr>
<td>Heffner et al., 1971</td>
<td>17, M</td>
<td>rt subfrontal, meningeal</td>
<td>brain scan &amp; angiogram</td>
<td>resection: STR (tumor), GTR &amp; XRT (AVM)</td>
<td>benign astrocytoma</td>
<td>improved</td>
</tr>
<tr>
<td>Crowell et al., 1975</td>
<td>17, M</td>
<td>rt temporal</td>
<td>brain scan, ventriculogram, angiogram (no shunting)</td>
<td>STR ( \times 2 )</td>
<td>oligodendroglioma</td>
<td>NA</td>
</tr>
<tr>
<td>Zuccarello et al., 1979</td>
<td>50, M</td>
<td>lt temporal</td>
<td>angiogram</td>
<td>NA</td>
<td>astrocytoma</td>
<td>NA</td>
</tr>
<tr>
<td>Ho &amp; Wolfe, 1981</td>
<td>63, F</td>
<td>lt thalamus/rt cingulate</td>
<td>NA†</td>
<td>NA</td>
<td>astrocytoma</td>
<td>NA</td>
</tr>
<tr>
<td>Licata et al., 1986</td>
<td>60, F</td>
<td>lt occipital/temporal</td>
<td>CT, angiogram</td>
<td>STR (tumor; AVM not treated)</td>
<td>GBM</td>
<td>died, Month 14</td>
</tr>
<tr>
<td>Martinez-Lage et al., 1986</td>
<td>43, M</td>
<td>rt parietal/ventricular</td>
<td>CT, angiogram</td>
<td>GTR &amp; XRT</td>
<td>oligodendroglioma</td>
<td>improved</td>
</tr>
<tr>
<td>Goodkin et al., 1990</td>
<td>9, F</td>
<td>lt parietotemporal</td>
<td>CT, angiogram</td>
<td>STR ( \times 2 )</td>
<td>anaplastic astrocytoma</td>
<td>NA</td>
</tr>
<tr>
<td>Malcolm et al., 1991</td>
<td>41, M</td>
<td>rt frontal</td>
<td>CT, angiogram</td>
<td>GTR &amp; XRT</td>
<td>astrocytoma</td>
<td>improved, Month 6</td>
</tr>
<tr>
<td>Lee et al., 1996</td>
<td>45, M</td>
<td>lt temporooccipital</td>
<td>CT, angiogram</td>
<td>GTR</td>
<td>pleomorphic xanthoastrocytoma</td>
<td>NA</td>
</tr>
<tr>
<td>Harris et al., 2000</td>
<td>57, M</td>
<td>rt temporoparietal</td>
<td>MRI, angiogram</td>
<td>open biopsy &amp; XRT (tumor), STR (AVM)</td>
<td>anaplastic astrocytoma</td>
<td>improved</td>
</tr>
<tr>
<td>Ziyal et al., 2004</td>
<td>58, M</td>
<td>rt temporoparietal</td>
<td>MRI, postop angiogram</td>
<td>GTR</td>
<td>high-grade glioma</td>
<td>died, Month 4</td>
</tr>
<tr>
<td>present study</td>
<td>55, F</td>
<td>rt parietal</td>
<td>MRI, angiogram</td>
<td>embolization &amp; GTR</td>
<td>anaplastic oligodendroglioma</td>
<td>died, Day 6</td>
</tr>
</tbody>
</table>

* GBM = glioblastoma multiforme; GTR = gross-total resection; NA = not available; STR = subtotal resection; XRT = radiation therapy.
† The AVM was diagnosed at autopsy.
Disclaimer
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References

Accepted January 4, 2008.
Address correspondence to: Jonathan L. Brisman, M.D., Cerebrovascular and Endovascular Neurosurgery, Winthrop University Hospital, Suite 128W, 100 Merrick Road, Rockville Centre, New York 11570. email: jbrisman@neurosurgeryli.com.