Pure sylvian fissure arteriovenous malformations

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Object. Pure sylvian fissure arteriovenous malformations (AVMs) are vascular malformations confined to the sylvian fissure without parenchymal involvement. Because the branches of the middle cerebral artery are arteries of passage and the margins between the AVM and the insula cortex may be ill defined, many surgeons regard pure sylvian fissure AVMs as inoperable. The authors reviewed their surgical experience with eight patients harboring pure sylvian fissure AVMs to determine the incidence of operative morbidity.

Methods. All eight patients experienced seizures, five (63%) had headaches, and three (38%) experienced hemorrhages. Preoperatively, six patients (75%) were normal neurologically and two (25%) had neurological deficits. Five (63%) of eight sylvian fissure AVMs were located in the dominant hemisphere. The size of the nidus ranged from 6 to 27 cm³ (mean 14 cm³).

Complete removal of the AVM was documented by postoperative angiography in every case. Seizures were reduced or eliminated and headaches were relieved in all affected patients. Transient neurological deficits, which included aphasia, short-term memory loss, and hemiparesis, occurred in four patients (50%). Within 3 months, all patients were functioning independently with no new neurological deficits. The status of two patients who had had preoperative neurological deficits improved postoperatively. Neuropsychological testing showed no new cognitive deficits.

Conclusions. With appreciation for transient instances of postoperative morbidity, the outcome was excellent in all patients. The authors thus advocate microsurgery as the primary treatment for pure sylvian fissure AVMs.

Key Words • arteriovenous malformation • insula cortex • sylvian fissure • surgery

Surgical removal is the preferred treatment for intracranial AVMs. The best treatment for unruptured deep-seated AVMs in critical locations is controversial. Arteriovenous malformations that involve the sylvian fissure are difficult to remove surgically because they are fed by MCA branches and the AVM is adjacent to the internal capsule and basal ganglia. Speech and memory may be affected when the AVM is located in the dominant hemisphere. Additionally, the medial border of the AVM is often poorly defined along the insula cortex and the surgical approach is oblique rather than perpendicular. This has led many surgeons to label these AVMs inoperable and to refer these patients for stereotactic radiation therapy.

In 1987, Sugita, et al., classified sylvian fissure AVMs into four subdivisions according to their locations within the sylvian fissure. In these authors’ series of 16 patients, most of the AVMs involved the frontal lobe (medial). Other sylvian AVMs were located in the temporal lobe (lateral) and in the internal capsule or basal ganglia (deep). Two (13%) of 16 AVMs were entirely within the sylvian fissure and were classified as pure sylvian fissure AVMs. Sugita, et al., removed these AVMs with a low incidence of morbidity, although most patients had experienced a major hemorrhage with significant preoperative neurological deficits.

The majority of patients in our series had unruptured AVMs. Seizure, hemorrhage, and headaches were the three indications for treatment. We reviewed our surgical experience with eight pure sylvian fissure AVMs treated from 1989 to 1996 (Fig. 1) to determine the incidence of operative morbidity and whether removal of sylvian AVMs was justified. Since 1996, two additional patients with pure sylvian fissure AVMs have undergone surgery with excellent results.

Clinical Material and Methods

Between July 1989 and July 1996, eight patients (four men and four women) ranging in age from 17 to 56 years (mean 32 years) underwent surgical removal of a pure sylvian fissure AVM. This group represents 5% of all AVMs surgically treated at the Mayfield Clinic and associated hospitals during this time period.
The most common presenting symptom was seizures. Seven (88%) of eight patients experienced complex partial seizures and one (13%) had generalized tonic–clonic seizures. In one patient a progressive left-sided weakness developed, but resolved when anticonvulsant medications were administered. Headaches were associated with seizures in five patients (63%) and three patients (38%) experienced hemorrhages. Neurological deficits caused by hemorrhage included expressive aphasia and left-sided hemiparesis in one patient and expressive aphasia and right-sided hemiparesis in another; the third patient was neurologically normal despite having had three hemorrhagic episodes (Table 1).

Diagnostic Studies

Cerebral angiography was performed in all patients to establish the diagnosis and to plan endovascular therapy. The patients underwent MR imaging for operative planning. Also performed were Wada tests to establish hemisphere dominance and memory. Intraoperative and postoperative angiography were performed to ensure complete removal of the AVM.

The volume of the AVM nidus ranged from 6 to 27 cm³ (mean 14 cm³). Five (63%) of the eight AVMs were located in the dominant sylvian fissure. In Case 1, the right hemisphere was dominant. The arterial supply primarily came from branches of the MCA. The anterior choroidal artery supplied the AVM in Cases 1, 4, and 8, and the posterior choroidal artery supplied the AVM in Cases 1 and 8. Venous drainage was predominantly cortical. Drainage through the basal vein of Rosenthal was present in two patients (Cases 1 and 8).

Neuropsychological testing was performed before and after surgery. All major domains of cognitive functioning were tested using standard clinical instruments. The battery of tests included general intelligence, verbal fluency, naming ability, memory processing, attention, abstract reasoning, manual motor skills, and psychosocial status.

Endovascular Therapy

Embolization was performed in Cases 3, 4, and 6. The other patients did not undergo embolization because vessels feeding their AVMs were too small to cannulate and the risk of occluding vessels of passage was too high. Amytal testing was performed before embolization. To embolize the AVMs we used N-butyl cyanoacrylate glue and a polyvinyl alcohol sponge. Although embolization did not significantly reduce arterial flow, the embolic materials served as useful intraoperative landmarks during surgical removal.

Surgical Treatment

Before surgery a No. 5 French femoral arterial sheath was placed to facilitate intraoperative angiography and a central venous catheter was placed for blood pressure control. Diuretic medications, hyperventilation, and cerebral brain protection were used to achieve brain relaxation. The patients were positioned supine with the ipsilateral shoulder elevated. The head was placed in a radiolucent three-point fixation device (Ohio Medical Instruments, Cincinnati, OH) and rotated 45˚ contralateral to the AVM.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (yrs), Sex</th>
<th>Preop Presentation</th>
<th>Hemorrhage</th>
<th>Nidus (cm³)/Hemisphere</th>
<th>Arterial Supply</th>
<th>Venous Drainage</th>
<th>Postop Results</th>
<th>AVM Outcome Score†</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>17, M</td>
<td>CPS, aphasia, hemi</td>
<td>yes</td>
<td>9/dominant MCA br, AChA, PChA</td>
<td>cortical; VR improved</td>
<td>improved</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>56, M</td>
<td>CPS, aphasia, hemi</td>
<td>yes</td>
<td>6/dominant MCA br</td>
<td>cortical</td>
<td>improved</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>37, F</td>
<td>CPS, HA</td>
<td>yes</td>
<td>20/dominant MCA br</td>
<td>cortical</td>
<td>normal</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>19, F</td>
<td>CPS, HA</td>
<td>no</td>
<td>23/dominant MCA br, AChA</td>
<td>cortical</td>
<td>normal</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>29, F</td>
<td>CPS, HA</td>
<td>no</td>
<td>6/nondominant MCA br</td>
<td>cortical</td>
<td>normal</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>52, M</td>
<td>CPS, HA</td>
<td>no</td>
<td>9/nondominant MCA br</td>
<td>cortical</td>
<td>normal</td>
<td>0</td>
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<tr>
<td>7</td>
<td>27, F</td>
<td>tonic–clonic seizures</td>
<td>no</td>
<td>10/dominant MCA br</td>
<td>cortical</td>
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<td>0</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>23, F</td>
<td>hemi, CPS, HA</td>
<td>no</td>
<td>27/nondominant MCA br, AChA, PChA</td>
<td>VR, SP sinus improved</td>
<td>mild hand weakness</td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>

* AChA = anterior choroidal artery; CPS = complex partial seizures; HA = headaches; hemi = hemiparesis; MCA br = middle cerebral artery branches; PChA = posterior choroidal artery; SP = sphenoparietal; VR = basal vein of Rosenthal.
† According to the Cincinnati AVM Outcome Scale (Table 2). The mean score ± standard deviation for this series was 0.62 ± 0.78.
Surgery was performed with the patient in a state of induced hypotension (mean arterial pressure 50–60 mm Hg) to reduce operative blood loss and facilitate surgical removal of the AVM.

A modified pterional craniotomy with caudal extension was performed (Fig. 2). The dura was opened to allow for wide exposure of the sylvian fissure. The arachnoid overlying the sylvian fissure was opened and the fissure was split along its entire length. Coagulation of the tiny feeding arteries from the MCA trunks was performed using an automatic bipolar electrocautery (CBC-1; Radionics, Randolph, MA). The energy level was set at 4 to 8 W and the impedance was monitored to avoid sticking of tissue to the bipolar forceps. Insulated nonstick nickel alloy bipolar forceps were used with two sets of 0.5-, 1-, 1.5-, and 2-mm forceps. Increasing the heat from the electrocautery reduced the effectiveness of the nonstick forceps. Therefore, a second set was used to remove the AVM while the first set cooled. Extensive arachnoid dissection of the draining veins was the key to mobilizing the pure sylvian AVMs. Temporal lobe retraction is more easily tolerated than retraction of the insula. Wide exposure of the sylvian fissure reduced the oblique angle of approach and provided a more perpendicular trajectory. Whereas most parenchymal AVMs are removed in circumferential dissection, pure sylvian AVMs are resected by means of an anteroposterior or posteroanterior approach. Meticulous hemostasis was required to maintain a clear field of dissection between the feeding arteries and the vessels en passage. The entire sylvian fissure was opened before removal of the AVM. The NC-4 microscope (Carl Zeiss Inc., Thornwood, NY) is well suited for removal of large sylvian AVMs because the headpiece can be rotated nearly 360° to visualize the entire sylvian fissure. The posterior sylvian fissure was opened using bayoneted forceps and microscissors as it extended medially and superiorly into the brain (Fig. 3). The myriad of short, tiny feeding arteries off the MCA branches were coagulated and cut as they entered the AVM. A large draining vein, which was often visible on the surface, was retracted with the AVM. Self-retaining microretractors (2 and 4 mm) were used for retraction of the AVM (Fig. 4). Retraction of the surrounding brain was minimized by wide splitting of the sylvian fissure. Coagulation of the coils led to shrinkage of the AVM mass and created space to remove the AVM. If venous drainage was posterior through cortical veins or the basal vein of Rosenthal, the AVM was removed in a proximal-to-distal direction along the sylvian fissure. If venous drainage was via the sphenoparietal sinus or deep middle cerebral veins, the AVM was removed from the distal sylvian fissure and rolled out anteriorly. Major venous drainage was preserved until the arterial supply was completely removed. Meticulous hemostasis of the operative bed and inspection of the skeletonized MCA branches was performed to avoid postoperative hemorrhage (Fig. 5). Induced hypotension was maintained for 48 to 72 hours after surgery.

Fig. 2. Operative photograph showing the surface of the brain in a patient harboring a pure sylvian fissure AVM.

Fig. 3. Drawing showing how wide splitting of the sylvian fissure exposes an extraparenchymal AVM. Reprinted with permission from the Mayfield Clinic.

Fig. 4. Drawing depicting how retraction of the AVM clearly exposes the deep arterial supply and minimizes retraction injury to surrounding brain. Reprinted with permission from the Mayfield Clinic.

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Outcome Analysis

Evaluation of outcome was based on the Cincinnati AVM Outcome Scale (Scores 0–8), which measures four independent but related factors. These outcome measures include: physical assessment, functional status, a comparison of preoperative and postoperative symptoms (headache, seizures, and cognition), and extent of AVM removal. By using the Cincinnati AVM Outcome Scale, the efficacy of embolization, stereotactic radiation therapy, and surgery for the treatment of AVMs can be determined (Table 2).

Results

Complete removal of the AVM was documented by postoperative angiography in each case. One patient (Case 5) required a second surgery for removal of residual malformation shown on the postoperative angiogram. The giant sylvian AVM in Case 8 was removed in two stages. Patients in whom the AVM was embolized underwent surgical removal within 1 week.

Surgical Outcome

Transient neurological deficits, including aphasia, short-term memory loss, and hemiparesis, occurred in four patients (50%). Seizures were reduced or eliminated and headaches were relieved in all affected patients. Two patients (Cases 1 and 2) who had preoperative neurological deficits improved postoperatively. Within 3 months, all patients were functioning independently with no new neurological deficits. All patients received a Glasgow Outcome Scale score of 5 and the mean Cincinnati AVM Outcome Scale score was 0.62 ± 0.78 (standard deviation; range 0–2).

Neuropsychological Outcome

Neuropsychological testing revealed no new cognitive deficits. Overall test results reflected an unimpaired level of functioning for these individuals. In general, the patients demonstrated no significant neuropsychological evidence of focal or diffuse impairment.

Discussion

In the past, surgical removal of unruptured sylvian fissure AVMs was not justified because of the expected high incidence of morbidity. Only patients who had experienced a hemorrhage with major neurological deficits were
candidates for surgery. Our success with unruptured pure sylvian AVMs has led us to adopt a more aggressive approach to treatment.

The key to removal of pure sylvian AVMs with a low incidence of morbidity is to open the sylvian fissure as widely as possible and to use retraction primarily on the AVM itself. The technical challenge is to operate at an oblique angle between the MCA branches. The feeding arteries are small and major blood loss is usually not encountered. To skeletonize the MCA branches with the large number of short, tiny feeding arteries requires patience. Normal and abnormal arteries must be examined from the beginning to the end of the sylvian fissure. To preserve MCA branches that course through the AVM, dissection should extend well beyond the posterior aspect of the AVM into the distal sylvian fissure. This wide-view perspective reduces the risk of occluding arteries of passage. The third and fourth divisions of the MCA follow the posterior sylvian fissure medially and superiorly deep into the brain. The microscope requires nearly 180° of rotation from the anterior sylvian fissure to the posterior sylvian fissure to expose and remove large sylvian AVMs.

Draining veins may obscure the AVM and must be mobilized and retracted with it. Embolization does not significantly reduce flow to sylvian AVMs; however, embolization may be used to obliterate pedicular or perinidal aneurysms, which are frequently the source of hemorrhage. In addition, orientation is difficult because of the oblique angles of the sylvian fissure; coils or sponges placed within the vessels provide useful landmarks to define the boundaries of the AVM. Preoperative MR images, especially coronal projections, will provide additional orientation. Induced hypotension throughout treatment and postoperatively reduces intraoperative blood loss and the risk of postoperative hemorrhage, respectively.

The advantages of surgery are immediate relief of headaches and excellent seizure control. The risk of hemorrhage is also eliminated. The major disadvantage is the high transient morbidity rate. After surgery, Sugita, et al.,16 reported that 13 (81%) of 16 patients experienced hemiplegia or hemiparesis. In our series, the transient morbidity rate was 50%. Patients should be counseled that they will likely experience weakness and/or have difficulty with speech. They will require rehabilitation and must be prepared psychologically to accept the neurological deficit. In most cases, these deficits begin to resolve within 1 week and are gone within 1 month.

There are several drawbacks to treating pure sylvian AVMs with stereotactic radiotherapy. First, the patient will continue to experience headaches and seizures until the AVM is thrombosed. The latency interval to obliteration after stereotactic radiotherapy may range from 1 to 3 years.1,3,6,8,11,14,15 Second, the nidus is frequently long and narrow and requires overlapping isodose centers, which may increase the risk of radiation injury. The risk of radiation injury is higher than normal when the AVM is adjacent to eloquent brain tissue, because the small lenticulostrate arteries leading to the internal capsule and basal ganglia are susceptible to radiation injury.6,17 When the nidus volume is large, obliteration rates decrease and complication rates increase after stereotactic radiotherapy.5,11 In our series, six (75%) of the patients had an AVM volume that exceeded 8 cm³. Multiple small doses of stereotactic radiation treatment may reduce the risk of radiation injury in large AVMs; however, the efficacy of fractionated radiotherapy has not been established. Moreover, repeated stereotactic radiotherapy has been associated with a 14% rate of complications.9

Another strategy is to reduce the AVM nidus through embolization to facilitate successful stereotactic radiation treatment;1,8 however, most of the arterial supply in pure sylvian AVMs is en passage and embolization would not significantly reduce the size of the AVM nidus. Embolization also poses significant risks; in large series, major morbidity rates ranged from 7 to 13% and mortality rates averaged 1.5%.8,18 Dawson, et al.,9 found that only two (29%) of seven large AVMs treated with combined embolization and stereotactic radiotherapy were cured. Gobin, et al.,7 reported a 65% obliteration rate in AVMs treated with embolization and stereotactic radiotherapy and a 12% recanalization rate. Pollock, et al.,14 also found a 7% recanalization rate in patients in whom stereotactic radiation treatment had failed. Therefore, stereotactic radiotherapy must treat the whole nidus, including the embolized portion of the AVM.

After stereotactic radiotherapy, the risk of rebleeding during the latency period that precedes AVM obliteration is unchanged from that of the natural history of the AVM (approximately 3–4% per year).5,12 Risk factors associated with rebleeding after stereotactic radiotherapy include: associated arterial aneurysm, venous aneurysm, venous outflow obstruction, periventricular location, prior embolization, and partial surgical removal.5 Morbidity and mortality rates from rebleeding are significant and should be factored into the decision to provide stereotactic radiotherapy.

In the hands of surgeons experienced in removing complex intracranial AVMs, surgical removal is the best treatment option for pure sylvian AVMs. Embolization and stereotactic radiotherapy are not well suited for treating sylvian AVMs. Stereotactic radiotherapy should not be used as validation for treatment because a qualified surgeon is unavailable. Instead, the patient should be referred

### Table 2

<table>
<thead>
<tr>
<th>Outcome Measure</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>physical assessment</td>
<td></td>
</tr>
<tr>
<td>neurological deficit</td>
<td>none 0, minor 1, major 2</td>
</tr>
<tr>
<td>functional assessment</td>
<td>independent w/o deficit 0, independent w/ minor deficit 1, dependent or vegetative 2</td>
</tr>
<tr>
<td>change in symptoms*</td>
<td>improved 0, unchanged 1, worse 2</td>
</tr>
<tr>
<td>angiographic obliteration</td>
<td>complete 0, incomplete 2</td>
</tr>
</tbody>
</table>

* Change in preoperative symptoms (headache, seizure, and/or cognition deficit) observed after surgery.
to a cerebrovascular center that has experience treating complex AVMs.

Conclusions
Pure sylvian AVMs can be resected safely by wide splitting of the sylvian fissure and by using retraction on the AVM to avoid injury to the surrounding brain. In this series, all patients affected by headaches and seizures improved postoperatively. Transient neurological deficits occurred in half of the patients, but within 3 months these patients were functioning independently with minor or no neurological deficits. A comparison of preoperative and postoperative neuropsychological testing showed no new cognitive deficits in the eight patients. We recommend surgery as the primary treatment for pure sylvian AVMs.

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References

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