Arteriovenous malformations of the lateral ventricle

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Nine cases with arteriovenous malformations (AVM's) predominantly involving the lateral ventricle are presented. All the AVM's were small, but caused intraventricular hemorrhage in eight cases. Only two patients had an intracerebral hemorrhage large enough to warrant evacuation. Eight patients were under the age of 40 years at the onset of their disease. Computerized tomography demonstrated intraventricular hemorrhage in eight patients, and after intravenous administration of contrast medium a small area of enhancement with dilated subependymal draining veins was seen in seven. The lateral ventricles were of normal size in seven cases, and only two patients required a shunting procedure. Angiography demonstrated that the lesion was an AVM in eight patients, and did not visualize the lesion in the ninth. One patient suffered a recurrent intraventricular hemorrhage when the AVM was demonstrated, although repeated angiography had failed to disclose a vascular lesion at his first intraventricular hemorrhage 14 months before.

All nine lesions were resected by microsurgical techniques, and the results were excellent in eight patients. Of four caudate lesions, three were resected through a frontal transcortical approach and the other was operated on through an anterior transcallosal approach; the results were excellent in three of these patients. Only one (Case 4) was left with neurological deficits; he had confusion and disorientation following a right frontal transcortical approach. Even in the dominant hemisphere, lesions in the head of the caudate nucleus could be safely resected by an anterior transcallosal approach. Two choroidal lesions located in the temporal horn and trigone on the dominant side were resected through a middle temporal gyrus approach, and three thalamic lesions through a posterior transcallosal approach, all with excellent results. In all cases the brain opening required was about the width of the retractor (maximum 2.0 cm, average 1.5 cm).

KEY WORDS arteriovenous malformation caudate nucleus cerebral angiography choroid plexus intraventricular hemorrhage lateral ventricle

Case Reports

Case 1

This 11-year-old right-handed boy experienced the sudden onset of severe headache and vomiting followed by unconsciousness. He was admitted to another hospital where a spinal tap disclosed grossly bloody cerebrospinal fluid (CSF), and he was referred for further evaluation. Neurological examination 10 days postictus demonstrated a very mild right hemiparesis and a stiff neck. Computerized tomography (CT) demonstrated a high-density area in the head of the left caudate nucleus with slight ventricular dilatation (Fig. 1 left upper and lower). Angiography revealed a small AVM involving the head of the caudate nucleus, fed by the lenticulostrate arteries and drained chiefly via the anterior caudate vein (Fig. 1 upper right).
The AVM was located subependymally in the head of the caudate nucleus, and was approached through a left frontal transcortical incision. Using a bipolar coagulator, a plane of dissection was achieved with some difficulty, as most of the feeding arteries ascended from below through the head of the caudate nucleus. The AVM was resected, and immediately the patient suffered a slight expressive aphasia; the mild right hemiparesis was unchanged. Over the course of a month he regained the full strength in his right arm and leg, and full recovery of speech. Postoperative angiography left some doubt about complete excision; however, follow-up angiography 1 year later demonstrated complete removal of the AVM (Fig. 1 lower right).

Case 4

This 37-year-old right-handed man presented with sudden severe headache and vomiting. Neurological examination revealed marked neck stiffness, and CT scanning demonstrated a high-density area in the head of the caudate nucleus on the right as well as ventricular hemorrhage. On referral 2 weeks following the hemorrhage, there was no neurological abnormality. Computed tomography scanning disclosed a small hemorrhage in the head of the caudate nucleus with moderate ventricular dilatation (Fig. 2 upper). Angiography revealed a small AVM in the head of the caudate nucleus (Fig. 2 lower).

The AVM was subependymal in location and was totally resected through a right frontal transcortical approach; complete removal of the AVM was confirmed by angiography. Immediately after surgery, the patient was alert but apathetic. A week later he was uncooperative and markedly disoriented to place, time, and person. A CT scan disclosed only a small low-density area in the white matter and mild ventricular dilatation. Neither placement of a right ventriculoperitoneal shunt nor psychiatric consultation relieved his organic mental symptoms, although the lateral ventricle became smaller. At follow-up review 6 months, 1 year, and 2 years later, he remained markedly confused, disoriented, and disinterested.

Case 5

This 35-year-old right-handed woman suddenly developed severe headache and a right hemiparesis. Neurological examination 4 days following the ictus demonstrated slight papilledema, a slight right hemiparesis with an extensor plantar response, and mild neck stiffness. A CT scan disclosed an intraventricular hemorrhage with a small high-density area in the head of the left caudate nucleus. Angiography demonstrated a small AVM in the head of the caudate nucleus, as seen in Cases 1 and 4. The AVM was located subependymally in the caudate nucleus, and was excised via an anterior transcallosal transventricular route. Postoperatively, the patient's neurological examination was normal except for the slight hemiparesis, which cleared completely in a month.

Case 6

This 33-year-old right-handed man had the acute
onset of neck and shoulder pain followed by severe generalized headache. The following day the headache became more severe and he was evaluated at an outside hospital, where a spinal tap demonstrated grossly bloody CSF. Neurological examination at our institution 4 days following the ictus disclosed a right central facial paresis, a stiff neck, and moderate disorientation to place and time. A CT scan demonstrated intraventricular hemorrhage, and angiography disclosed an AVM in the temporal horn and trigone of the left lateral ventricle, supplied by the anterior and posterior choroidal arteries. Venous drainage was via an inferior ventricular wall vein (Fig. 3 left). The AVM was located in the choroid plexus, and was easily resected through a left temporal craniotomy and middle temporal gyrus approach. Following resection, the patient had a slight acalculia, finger agnosia, confusion of laterality, and a right homonymous hemianopsia, in addition to the preoperative findings. In the course of 2 weeks, the Gerstmann syndrome and the other signs had completely resolved, and the patient returned to his previous work. Angiography confirmed complete resection of the AVM (Fig. 3 right pair).

Case 7

This 7-year-old right-handed boy presented with a sudden severe headache. He was evaluated at an outside hospital, where CT demonstrated intraventricular hemorrhage (Fig. 4 left) and angiography disclosed a small AVM in the dorsal aspect of the left thalamus, supplied by the posterior choroidal arteries and drained by the thalamostriate vein (Fig. 4 upper right). The AVM was located subependymally in the dorsal surface of the thalamus, but was not related to the choroid plexus. The lesion was resected through a left parieto-occipital craniotomy via the posterior transcoccal route. Some
feeding vessels, running superficially from posteriorly, were easily coagulated. A plane of dissection was achieved with some difficulty, because some of the feeding arteries ascended through the thalamus. Following resection of the AVM, the patient was confused and apathetic for 4 days, and diffident and reticent for another 4 days. His condition then improved. Follow-up neurological and neuropsychological examinations were normal. Angiography demonstrated complete resection of the AVM (Fig. 4 lower right), and CT demonstrated a small low-density area in the dorsal aspect of the thalamus.

Case 8
This 23-year-old right-handed woman had the sudden onset of double vision, numbness in the left upper limb, weakness of the left lower limb, and gradual drowsiness 4 days prior to admission. She was first evaluated at an outside hospital, complaining of severe headache and frequent vomiting. A CT scan demonstrated a hemorrhagic lesion in the right thalamus, extending downward to the midbrain; dilatation of the lateral ventricles was also revealed (Fig. 5 upper). A cavum vergae was also seen.

On referral the patient was drowsy, with bilateral
Arteriovenous malformations of the lateral ventricle

low-density area remaining in the left subthalamus (Fig. 5 lower).

Summary of Cases

Table 1 provides clinical details of the nine patients in this series. The age at onset ranged from 7 to 51 years, and only one patient was over the age of 40 years. The presenting symptom was intraventricular hemorrhage in eight, with a small intracerebral hemorrhage in two. Only two patients (Cases 3 and 8) had an intracerebral hemorrhage large enough to be evacuated. Contrast-enhanced CT scans demonstrated a small area of enhancement, with a dilated subependymal draining vein in seven cases. The ventricles were of normal size in eight patients. Eight lesions were demonstrated by angiography; in only one (Case 8) was the AVM not visualized angiographically. In one patient (Case 9) repeated angiography had failed to demonstrate an earlier intraventricular hemorrhage, and 14 months later, when the hemorrhage recurred, the AVM was discovered in the dorsal surface of the thalamus.

The AVM's were less than 2 cm in diameter in all patients. Four were located in the head of the caudate nucleus (in the anterior horn), two in the choroid plexus (in the temporal horn and trigone), and three in the dorsal surface of the thalamus (in the trigone); five were on the left side and four on the right. The AVM's were resected at a single-stage operation with microsurgical techniques, and total removal of the lesion was verified by angiography and CT scanning. The results were excellent in eight and persistent neuropsychological deficits occurred in only one patient (Case 4). In all patients, the size of brain opening and leukotomy required was approximately the width of the blade of a retractor, maximum 2 cm, average 1.5 cm.

Discussion

Arteriovenous malformations (AVM's) involving the lateral ventricle and basal ganglia comprise from 4% to 10% of all cerebral AVM's. They occurred in 8% of the 50 children reported by Harwood-Nash and Fitz. In a series of 56 cases presented by Gerosa, et al., these deep lesions were even more common in children; nine (16%) in the basal ganglia, seven (13%) in the trigone, and three (5%) in the thalamus. The small cryptic AVM's, angiographically invisible but detectable on CT scanning, are also present in the thalamus and caudate nucleus.

Luessenhop and Gennarelli have formulated an anatomical grading scheme to correspond to the number of tertiary arteries directly supplying AVM's and the degree of surgical difficulty involved in total excision; Grade I lesions can be operated on without morbidity and mortality regardless of clinical state and location, and Grade IV AVM's are probably not operable. The great majority of AVM's supplied by the lenticulostriate arteries are described as Grade IV, and operable periventricular AVM's are those supplied by...
TABLE 1

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Location of AVM</th>
<th>Presentation</th>
<th>Preop Status</th>
<th>Operative Approach</th>
<th>Postop Status</th>
<th>Follow-Up Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>11, M</td>
<td>Lt head of caudate nucl</td>
<td>IVH</td>
<td>mild rt hemiparesis</td>
<td>lt frontal transcortical</td>
<td>slight expressive aphasia, mild hemiparesis unchanged</td>
<td>completely recovered</td>
</tr>
<tr>
<td>2</td>
<td>51, F</td>
<td>Lt choroid plexus</td>
<td>IVH</td>
<td>normal</td>
<td>lt middle temporal gyrus</td>
<td>normal</td>
<td>healthy</td>
</tr>
<tr>
<td>3</td>
<td>18, M</td>
<td>rt head of caudate nucl</td>
<td>IVH, ICH</td>
<td>lethargic, lt hemiparesis</td>
<td>rt frontal transcortical</td>
<td>mild lt upper extremity weakness</td>
<td>completely recovered</td>
</tr>
<tr>
<td>4</td>
<td>37, M</td>
<td>rt head of caudate nucl</td>
<td>IVH, ICH</td>
<td>normal</td>
<td>rt frontal transcortical</td>
<td>confusion, disorientation</td>
<td>no change, unemployed</td>
</tr>
<tr>
<td>5</td>
<td>35, F</td>
<td>Lt head of caudate nucl</td>
<td>IVH, ICH</td>
<td>mild rt hemiparesis</td>
<td>anterior transcallosal</td>
<td>no change</td>
<td>completely recovered</td>
</tr>
<tr>
<td>6</td>
<td>33, M</td>
<td>Lt choroid plexus</td>
<td>IVH</td>
<td>normal</td>
<td>lt middle temporal gyrus</td>
<td>slight Gerstmann syndrome</td>
<td>completely recovered</td>
</tr>
<tr>
<td>7</td>
<td>7, M</td>
<td>Lt thalamus</td>
<td>IVH</td>
<td>normal</td>
<td>posterior transcallosal</td>
<td>normal</td>
<td>healthy</td>
</tr>
<tr>
<td>8</td>
<td>23, F</td>
<td>Rt thalamus</td>
<td>ICH</td>
<td>lethargic, rt 3rd nerve palsy, bilat MLF lesions, loss of brisk tendon reflexes, dense lt hemiparesis</td>
<td>posterior transcallosal</td>
<td>dense lt hemiparesis</td>
<td>no change</td>
</tr>
<tr>
<td>9</td>
<td>39, M</td>
<td>Rt thalamus</td>
<td>recurrent IVH</td>
<td>normal</td>
<td>posterior transcallosal</td>
<td>normal</td>
<td>healthy</td>
</tr>
</tbody>
</table>

* Abbreviations: AVM = arteriovenous malformation; nucl = nucleus; IVH = intraventricular hemorrhage; ICH = intracerebral hemorrhage; MLF = medial longitudinal fascicle.

Surgical approaches are available that leave no neuropsychological sequelae, or at least reduce the possibility of sequelae.

The AVM's located in the head of the caudate nucleus in the nondominant hemisphere can be resected via a frontal transcortical or an anterior transcallosal route without neuropsychological sequelae. For those located in the dominant hemisphere, an anterior transcallosal approach is preferable since frontal cortical incisions in the dominant hemisphere are not without complications. Although a frontal transcortical approach may give excellent results without sequelae, it can result in neuropsychological deficits. In the patient reported by Carton and Hickey transient mental confusion, poor memory, tendency to perseverate, and emotional lability were observed immediately postoperatively, and there was minimal reduction in performance on psychological testing at 15-month follow-up review. Ralston and Papatheodorou described postoperative neurological findings including limitation of upward gaze and slight difficulty in naming objects. As mentioned above, frontal cortical incisions are not without their complications, even in the nondominant hemisphere, as shown in our Case 4. We believe that lesions in the head of the caudate nucleus should be resected through an anterior transcallosal approach as recommended by Shucart and Stein.

The AVM's in the temporal horn and trigone on the nondominant side can be approached and resected one or two of these arteries. On the other hand, AVM's of the choroid plexus can be considered as Grade III. For Grade III lesions, location and clinical state are probably more important than anatomical grading. One of the studies on the natural history of cerebral AVM's has shown that average yearly risk for first hemorrhage is between 2% and 3%, the risk of rebleeding is 6% in the 1st year, and thereafter the average rebleeding rate is 2% per year for up to 20 years. Luessenhop and Rosa, taking into consideration this study and equating another grading scheme to the maximum measured diameter of AVM's in the lateral angiograms, have concluded that for Grade I (up to 2 cm) and Grade II (2 to 4 cm) cerebral AVM's, the risk involved in nonsurgical therapy exceeds that of surgical treatment, so excision is indicated; however, for Grade III (4 to 6 cm) and more extensive AVM's, the surgical risk probably exceeds the natural risk in most circumstances for patients after the fourth to fifth decade of life.

The risk of initial bleeding appears to be greater with small lesions, as seen in our limited series. Small AVM's predominantly involving the lateral ventricle can be resected without deficits on the nondominant hemisphere and without or with minimal and acceptable deficits on the dominant side.

There are three main locations of AVM's in the lateral ventricle; anterior horn (the head of the caudate nucleus), trigone and temporal horn (the choroid plexus), and trigone (the dorsal surface of the thalamus).
Arteriovenous malformations of the lateral ventricle

without a field defect by a short transverse incision in the superior or middle temporal gyrus carried down in the trigone.\textsuperscript{3,26} On the dominant side the lesions may be approached transcortically\textsuperscript{10,12,26,29,30} or transcallosally through the splenium or just in front of it,\textsuperscript{3,26} with or without significant sequelae.

Thalamic AVM's presenting on the ventricular surface may be resected via a prefrontal, temporal, or transcallosal approach.\textsuperscript{3,12} When the lesions are small or medium-sized and receive blood supply from both the anterior and posterior choroidal arteries on the nondominant side, they can be resected via a temporal route if the cortical incision is made somewhat wider. We believe that it is better to approach lesions on the dominant side via a posterior transcallosal route or, as an alternative, via both the temporal and callosal routes.\textsuperscript{26} The temporal route provides a better way to occlude the feeding vessels from the anterior choroidal artery, while the posterior transcallosal route facilitates occlusion of the feeding vessels from the posterior choroidal arteries. A large temporal incision may give good access to both the anterior and posterior choroidal arteries; however, this is not desirable on the dominant side.

Wharen, \textit{et al.},\textsuperscript{29} operated on a right-handed girl with a large AVM involving the left thalamus and glomus of the choroid plexus; they approached the lesion via a left parieto-occipital craniotomy by a leukotomy in the medial portion of the left parietal lobe, entering the atrium of the lateral ventricle. Immediately following surgery, the patient had an expressive aphasia, right homonymous hemianopsia, and a flaccid right hemiparesis. Three months later she could walk independently, but the right arm was still very weak; her speech is conversational, with some deficits in auditory processing and writing. Left trigonal\textsuperscript{10} and thalamic\textsuperscript{30} AVM's have been exposed and resected by a high parieto-occipital approach across the trigone of the lateral ventricle, with good results. Two patients reported by Wilson, \textit{et al.},\textsuperscript{30} were temporarily worse but then improved: one returned to the preoperative level of impairment and the condition of the other was ameliorated.

With parapependuncular AVM's or those located medial to the trigone of the lateral ventricle, Stein\textsuperscript{26} has recommended that the approach be transcortical via the posterior middle temporal gyrus, and if the removal is difficult via this route, that the resection be done in two stages. The first stage of this procedure approaches via the posterior infratemporal route and the second stage via an interhemispheric parafalx exposure to remove the residual portion of the AVM's.

In the series reported by Juhász,\textsuperscript{12} one AVM located in the corpus striatum, the genu of the internal capsule, anterior part of the thalamus, and in centrum semiovale on the dominant side was supplied by the anterior and posterior choroidal and lenticulostriate arteries. It was removed via two cortical incisions, a parietal one for the feeding vessels from the posterior choroidal arteries when a portion of the AVM was removed, and a frontal one where the residual AVM was removed with considerable bleeding. Three years after the operation the patient had a slight motor aphasia. Another AVM located in the nondominant temporal horn was re- moved at that time without any neurological deficits after resection of the right laterobasal portion of the temporal lobe.

Various approaches have been proposed for meningiomas of the lateral ventricle involving the trigone on the dominant side.\textsuperscript{4,11,25} Approaches through the inferior parietal lobule or the temporal lobe very often cause transient or permanent speech, motor, or sensory deficits, in addition to the damage to the optic radiation. An approach via the posterior portion of the corpus callosum has been described as safe by Kempe and Blaylock,\textsuperscript{13} although a disconnection syndrome with alexia without agraphia has been reported.\textsuperscript{15} A para- median parieto-occipital approach may carry the risk of damage to the higher function of the parietal lobe.\textsuperscript{6,11,13} We believe that a posterior transcallosal approach, although it sacrifices a functionally significant portion of the corpus callosum, gives a better and less traumatic route for removal of AVM's and meningiomas located in the trigone. The neuropsychological sequelae are minimal and acceptable when the incision in the corpus callosum or splenium is small.\textsuperscript{3,6,11,13}

When the AVM's are large and there is need to split considerable amount of the corpus callosum, we do not know whether a temporal, high parietal, or posterior transcallosal approach is better or more desirable. Large AVM's involving the lateral ventricle in the dominant hemisphere may be unsuitable for a direct operative procedure, and it may be better to treat them with intravascular techniques.

In the presence of sizable intracerebral hematoma, we find that it is wise to evacuate the hematoma to reduce intracranial pressure and save the life of the patient first. The AVM can then be resected with a microsurgical technique after the patient's condition has stabilized.\textsuperscript{23,27}

References


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