Angiographically occult arteriovenous malformations

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Eight cases of histopathologically proven arteriovenous malformations (AVM's) which were not visualized on angiography are presented. As is typical with these lesions, most of the patients in this series presented with hemorrhage, seizures, or episodic or progressive neurological symptoms suggestive of a neoplasm. The diagnosis of angiographically occult AVM was highly suspected preoperatively in each case based on the combination of computerized tomography (CT) and magnetic resonance (MR) findings. The CT scans in all cases showed moderately hyperdense lesions which enhanced mildly or moderately in a nonhomogeneous pattern with administration of contrast material. The MR image showed one or more bright areas interspersed with areas of low or absent signal peripherally or centrally on both T1- and T2-weighted images. The AVM was totally excised in seven patients and partially excised in one patient, with favorable results in all. The clinical management and differential diagnosis of angiographically occult AVM's are discussed. In patients with a clinical course and radiological studies suggestive of an occult AVM, removal of the lesion, if accessible, should be performed in order to rule out a neoplasm and prevent subsequent hemorrhage and progression of symptoms.

Key Words: arteriovenous malformation · cerebral angiography · cryptic arteriovenous malformation · occult arteriovenous malformation

The existence of vascular malformations of the brain which are not visualized on angiography has been recognized for many years. These "occult" or "cryptic" lesions have usually been diagnosed pathologically after surgical removal or at autopsy. Computerized tomography (CT) has helped in the diagnosis of angiographically occult arteriovenous malformations (AVM's). Although CT scans can suggest the diagnosis of AVM's occult to angiography, the differential diagnosis of these lesions includes intra-axial neoplasm, granulomatous lesions, and even a vascular meningioma. Magnetic resonance (MR) imaging has allowed increased confidence in suspecting the correct diagnosis of angiographically occult AVM's. However, it should be noted that although the MR image may suggest an occult AVM, some neoplasms can show an identical MR appearance.

Cavernous angiomas can also be occult angiographically and present a CT and MR appearance that is indistinguishable from that of occult AVM's, although frequently cavernous angiomas are larger. Patients with cavernous angiomas frequently present with a slowly progressive clinical deficit at times punctuated by episodic worsening. Venous angiomas have a more characteristic CT and MR appearance (normal on plain CT scan, linear density on enhanced CT, and a tubular signal-void area on T2-weighted MR imaging) and are usually, although not always, seen in the late venous phase of the angiogram. Therefore, they should not be confused with occult AVM's. It is particularly important to avoid this confusion since, in general, venous angiomas are very benign lesions that should be left alone.

This is a report of eight patients with angiographically occult AVM's in which the diagnosis was confirmed pathologically. Six of these patients have been presented before in a report concerning the radiographic and, particularly, the MR characteristics of these lesions. The patients often had similar presenting symptoms, which were episodic or progressive and occasionally suggested a neoplasm. The cases in the current report confirm the concept that AVM's fail to appear angiographically due to thrombosis, their small size, or pressure from a hematoma.

Clinical Material and Methods

The clinical, surgical, and histological features of eight patients with angiographically occult AVM's are
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FIG. 1. Radiograms in Case 2. A: Contrast-enhanced computerized tomography (CT) scan showing irregular enhancement of the lesion located in the left brachium pontis. Enhancement in and adjacent to the lesion (arrow) probably represents small vessels. B: Unenhanced CT scan showing a heterogeneous, largely hyperdense lesion. C: Magnetic resonance (MR) image performed 2 days after the CT scan. There is a homogeneous bright signal of short T1 which is consistent with nonacute hemorrhage. The areas of lower signal seen more centrally (arrow) are consistent with hemorrhage and do not correspond to areas of enhancement (vessels) seen on CT scanning. D: A T2-weighted MR image of a slice 7.5 mm above the image shown in C. There are bright “islands” of increased T2 signal with a boundary zone of absent or reduced signal between the “islands.”

presented in Table 1. There were five men and three women, ranging in age from 18 to 41 years. The clinical symptoms were often episodic and occurred over weeks, months, or years. The symptoms usually included headache with or without nausea and vomiting, focal or grand mal seizures, and focal neurological deficits. In Case 7 a focal seizure of the right upper extremity was the presenting symptom. Three patients suffered hemorrhage subsequent to a normal angiogram. A CT scan without and with contrast agent (using GE 8800 and 9800 scanners) was the initial study obtained in all of the patients. Selective catheter angiography with magnification and subtraction was obtained before or after MR imaging. The angiograms were reviewed retrospectively and failed to demonstrate any intrinsic vascular shunting or abnormal vessels; in some patients with a hemorrhage, a mass effect was demonstrated. Magnetic resonance imaging was performed using a previously described technique.26,27 Surgical exploration was undertaken when the CT and MR data in conjunction with the clinical presentation were indicative of an angiographically occult AVM.

Results

Radiological Studies

There was no evidence of pathological vessels or arteriovenous shunting on careful retrospective study of the angiograms. Mass effect was apparent in two of the cases and not in the remaining six cases. The size of the lesions on the CT scans was less than 3 cm in all of the patients. In only two patients (Cases 3 and 6) was calcification definitely identified; in Cases 1, 2, and 4 there was heterogeneous hyperdensity which suggested the possibility of calcification (Fig. 1B). In Case 1 the original plain CT showed focal acute hemorrhage in the left temporal lobe with slight enhancement at the periphery of the hematoma with administration of contrast material. There were focal areas of enhancement consistent with small blood vessels. A plain CT scan obtained 2½ months after the initial study revealed a heterogeneous hyperdense lesion; moderate heterogeneous enhancement was shown on the contrast study. In Case 5 the lesion initially had an isodense appearance on plain CT. Four months later, CT showed the lesion with low mixed density in the center and an irregular thick ring consistent with recent hemorrhage. This appearance raised the possibility of hemorrhage into a neoplasm as opposed to a vascular malformation. Arcuate foci of enhancement representing small abnormal blood vessels were seen on the contrast CT scans in Cases 1, 2, and 4 (Fig. 1A).

The T1-weighted MR image in Cases 1, 2, 3, and 5 revealed one or more foci of short T1 signal which appeared as bright islands within the lesion (Fig. 1C). Cases 4, 6, 7, and 8 showed only a low signal. In addition, areas of very low or absent signal on T2-weighted images were present peripherally or centrally in four cases (Fig. 1C). On T2-weighted images one or more foci of long T2 signal, appearing as bright islands, were seen in five cases (Fig. 1D). This was accompanied by a very low or absent T2 signal which was arranged peripherally in four cases, and peripherally plus centrally in one case (Fig. 1D). In Cases 6 and 8 the entire area showed a low to absent T2 signal.

Surgery

The lesion identified on CT or MR imaging was totally excised using microsurgical techniques in seven of the eight cases. In Case 3 only partial excision of the lesion was possible. All of the lesions had gross evidence of old hemorrhage at surgery. In Case 6 there was evidence of both old and recent hemorrhage. The appearance of a thrombosed AVM was noted in Cases 2.
described two incidental lesions discovered at autopsy, these, two had venous malformations, one a cavernous angioma, and the fourth a tiny AVM. The authors also described small vascular malformations in six patients at autopsy. Four patients suffered fatal hemorrhages; of these, two had venous malformations, one a cavernous angioma, and the fourth a tiny AVM. The authors also described two incidental lesions discovered at autopsy, one of which was a telangiectasia and the other a cavernous angioma. Subsequent to this report there have been many descriptions of histologically verified angiographically occult vascular malformations. The pathology of the lesions has included telangiectasia, cavernous angioma, venous angioma, and true AVM's. As more cases have been reported, the term "cryptic" or "occult" cerebrovascular malformation has been used to describe any vascular malformation which cannot be detected angiographically. The eight lesions described in this study are true AVM's, separated from other occult malformations for descriptive purposes, although at times the difference between true AVM's, telangiectasias, and venous angiomas cannot be definitively established histologically and it appears that "mixed" or "transitional" histological forms do occur.

The AVM's occult to angiography consist of two main types of lesions. One group of patients harbor a typical AVM which fails to be visualized angiographically because of its small overall size, the small caliber of its component vessels, or slow blood flow through the lesion. These lesions may bleed repeatedly and can cause significant neurological deficits. It is this type of small lesion that was initially described by Margolis, et al., in association with fatal hemorrhage. Wakai, et al., reported 13 cases of angiographically occult AVM's where the hemorrhage was often large and devastating. These authors described the microsurgical technique recommended for exploring the hematoma cavity. Briefly, a small corticotomy is made over the thinnest portion of the cortex covering the hematoma. The brain is retracted and the hematoma is evacuated in layers in order to progressively separate the clot from the cavity wall. Under the microscope, the wall of

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex, Age (yrs)</th>
<th>Site of Lesion</th>
<th>Clinical Presentation</th>
<th>Gross Appearance at Surgery</th>
<th>Histological Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M, 26</td>
<td>lt posterior temporal</td>
<td>3 mos episodic headache, nausea &amp; vomiting, difficulty with reading</td>
<td>encapsulated mass, old clot, gliosis</td>
<td>vascular malformation</td>
</tr>
<tr>
<td>2</td>
<td>M, 41</td>
<td>lt pontocerebellar</td>
<td>1½ mos episodic instability, dizziness, nausea, visual blurring</td>
<td>tangle of vessels, some thrombosed; gliosis, hematoma</td>
<td>AVM</td>
</tr>
<tr>
<td>3</td>
<td>M, 27</td>
<td>posterior ponto-mesencephalic</td>
<td>36 mos episodic blurred vision, diplopia, headache, speech difficulty</td>
<td>encased mass within old hemorrhage, thrombosed vessels</td>
<td>consistent with vascular malformation</td>
</tr>
<tr>
<td>4</td>
<td>M, 40</td>
<td>lt fourth ventricular</td>
<td>12 mos episodic vertigo, nausea &amp; vomiting, headache, diplopia, SAH</td>
<td>small mass, old hemorrhage with thrombosed vessels</td>
<td>AVM</td>
</tr>
<tr>
<td>5</td>
<td>F, 31</td>
<td>lt frontal</td>
<td>8 mos episodic minor seizures, recent generalized seizures</td>
<td>encased mass containing old hemorrhage, gliosis</td>
<td>AVM</td>
</tr>
<tr>
<td>6</td>
<td>F, 40</td>
<td>lt frontal</td>
<td>many years of lt frontal headache, 16 mos earlier acute headache, nausea &amp; vomiting, then obtundation &amp; grand mal seizures</td>
<td>localized mass, old &amp; recent hemorrhage, gliosis</td>
<td>AVM</td>
</tr>
<tr>
<td>7</td>
<td>M, 18</td>
<td>lt Rolandic</td>
<td>focal seizure, with numbness of right side of face, arm &amp; chest</td>
<td>small-vessel AVM with no large feeding vessels, low flow pressure, partial thrombosis</td>
<td>AVM</td>
</tr>
<tr>
<td>8</td>
<td>F, 23</td>
<td>rt temporal</td>
<td>partial complex seizures</td>
<td>compact lesion with small arteries, venules patent but under low pressure</td>
<td>AVM</td>
</tr>
</tbody>
</table>

4, 7, and 8. The remainder of the lesions appeared grossly as vascular malformations with fine, delicate tangles of vessels usually under relatively low pressure. The lesions in Cases 2, 7, and 8 were thought to be completely thrombosed preoperatively. At surgery these lesions proved to be only partially thrombosed with some residual patent vessels. These vessels were arterialized and were under relatively low pressure. All of the lesions were well demarcated with surrounding gliosis. All of the patients did well postoperatively, and no morbidity resulted from the procedure. The follow-up period has ranged from 6 months to several years and, to our knowledge, no patient has had a subsequent hemorrhage or has suffered subsequent deterioration.

The excised lesions were carefully reviewed in an attempt to identify the exact type of vascular malformation present. The diagnosis of AVM was made with confidence in all but two cases. The lesion in Case 1 was totally excised, but only fragments were available for histological examination. These fragments demonstrated abnormal vessels consistent with AVM. The lesion in Case 3 was partially excised; it was associated with abnormal vessels consistent with a vascular malformation, most likely an AVM.

**Discussion**

The term "cryptic" or "occult" vascular malformation has been used for many years. Margolis, et al., described small vascular malformations in six patients at autopsy. Four patients suffered fatal hemorrhages; of these, two had venous malformations, one a cavernous angioma, and the fourth a tiny AVM. The authors also described two incidental lesions discovered at autopsy, one of which was a telangiectasia and the other a cavernous angioma. Subsequent to this report there have been many descriptions of histologically verified angiographically occult vascular malformations. The pathology of the lesions has included telangiectasia, cavernous angioma, venous angioma, and true AVM's. As more cases have been reported, the term "cryptic" or "occult" cerebrovascular malformation has been used to describe any vascular malformation which cannot be detected angiographically. The eight lesions described in this study are true AVM's, separated from other occult malformations for descriptive purposes, although at times the difference between true AVM's, telangiectasias, and venous angiomas cannot be definitively established histologically and it appears that "mixed" or "transitional" histological forms do occur.

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*AVM = arteriovenous malformation; SAH = subarachnoid hemorrhage.
cavity can be inspected to locate and remove a small AVM, usually in the wall of the hematoma cavity. The hematoma cavity usually has a region of surrounding gliosis which determines the plane of dissection.

In the second group of patients, angiography fails to visualize the AVM because of thrombosis or compression by the pressure of a hematoma. Completely thrombosed AVM's most frequently present with seizures. It is interesting to note that, in a report of seven cases of thrombosed AVM's by Wharen, et al., only one lesion was partially thrombosed. Cases 2, 7, and 8 in our series had partially thrombosed lesions. In these lesions there was pulsatile flow under relatively low pressure. One likely reason for AVM thrombosis is thought to be previous hemorrhage. In all of our cases there was evidence of old hemorrhage of various ages. This finding is similar to that of Wharen, et al., in which six of their seven lesions had histopathological evidence of previous subclinical hemorrhage. The risk of repeat hemorrhage justifies removal of these lesions whenever possible. Regarding hemorrhage, Wakai, et al., reviewed the literature and determined that between 27% and 53% of patients with a lobar hemorrhage and normal early angiogram will prove to have an AVM at surgery or with subsequent angiography. This figure excludes patients with hypertension and elderly patients likely to have amyloid angiopathy. Occurrence of previous hemorrhage in part helps to explain the CT and MR appearance of these lesions.

The literature describing the radiological features of lesions occult to angiography is somewhat confusing in that it includes lesions that on pathological examination are true AVM's, venous malformations, telangiectasias, or cavernous angiomas. On plain CT these various lesions are generally described as hyperdense or isodense; they are usually less than 3 cm in diameter and have little or no surrounding edema. There are often punctate areas of hyperdense foci indicative of calcification. With the administration of a contrast agent there is only mild to moderate enhancement of the lesions and, rarely, there is no enhancement. Small areas of intense enhancement may be observed which represent contrast medium in small blood vessels. It has been suggested that lesions showing minimal or no enhancement on contrast CT scans suggest the diagnosis of a cavernous angioma as opposed to other types of occult vascular malformations where definite contrast accumulation is seen. The CT findings described in the present study are similar to those described in a recent report of angiographically occult AVM's in childhood.

The MR image of these patients can often help to suggest the diagnosis of angiographically occult vascular malformations with increased confidence. Our eight patients had MR scans that were similar in appearance to those described previously for occult AVM's. There are often circumscribed regions of low intensity, most prominent on T2-weighted images. These areas are interspersed with foci of mixed signal intensity patterns ("islands") which correspond to different stages of evolution within the hematoma. The low-intensity or signal-void areas most likely represent hemosiderin deposits, which diminish the signal because of the magnetic susceptibility effect of hemosiderin. Other less likely possibilities for this appearance include blood flow within the lesion, calcification, or a draining vein. Magnetic resonance imaging helped to suggest the correct diagnosis in our eight patients who presented with episodic chronic symptoms suggestive of a neoplasm. The CT scan and the MR image appear to be about equally sensitive in the detection of these lesions, although the MR image is probably slightly more specific. However, it should be emphasized that neither the radiological appearance nor the clinical history, alone or in combination, is 100% specific for this entity. We have seen one patient with three small hemorrhages over a 15-year period and another with two small hemorrhages over an 8-year period; both of these patients had angiographically occult lesions and a CT and MR appearance characteristic of an occult AVM, and at surgery both proved to have low-grade astrocytomas.

Headaches and seizure disorders are the most common clinical presentation of angiographically occult AVM's. In addition, many of the patients present with episodic progressive worsening of a focal neurological syndrome. This presentation, in conjunction with the CT and MR findings, may suggest the presence of a low-grade neoplasm. There is at least one report where lesions in the thalamus were empirically irradiated on the assumption that they were gliomas, and at autopsy thrombosed AVM's were discovered. As emphasized above, although the MR appearance may be very suggestive of an AVM, surgery is still needed to establish the diagnosis and to eliminate the risk of future hemorrhage. An operation should be considered whenever the lesion is surgically accessible.

It is not clear what to do with inaccessible lesions such as those deep within the thalamus or brain stem. If the symptoms are mild and nonprogressive, or if the patient has had only one hemorrhage, it appears prudent to manage the patient conservatively with follow-up CT and MR studies at regular intervals. With progressive symptomatology or repeated hemorrhages, surgical resection may be considered since these lesions result in marked gliosis in the surrounding brain which offers a relatively safe plane of resection. Alternatively, blind irradiation, perhaps by the proton beam method, can be considered. Because of the fear of hemorrhage, closed stereotaxic biopsy is not generally recommended when this lesion is suspected. However, Daumas-Duport, et al., have reported a series of 14 patients who underwent stereotaxic biopsy of presumed angiographically occult AVM's in deeply situated areas of the brain. Five of these lesions proved to be cavernous angiomas, two capillary telangiectasias, three venous angiomas, two true AVM's, and one a vascu-
cular hematoma. Partial hematoma evacuation was performed in four patients. Five of the patients biopsied had “mild and transitory” aggravation of a preexisting motor deficit which had resolved in all but one patient by the time of discharge from the hospital.\(^7\)

In patients with an intracranial hemorrhage of unknown etiology and a normal initial angiogram, it is recommended that angiography be repeated 6 to 12 weeks after the hemorrhage, depending on the neurological condition of the patient. If the follow-up angiogram is again negative, yet the CT scan or MR image suggests an AVM, then it is reasonable to proceed with exploration, and excision if a lesion is found. A conservative approach is indicated if neither study is suggestive of an occult AVM.

In summary, angiographically occult AVM’s can be suspected with increased (although not yet absolute) confidence using CT and MR criteria. These lesions warrant surgical exploration and removal in order to confirm the diagnosis and prevent recurrent hemorrhage. The eight patients reported in the current series have done well clinically after total excision of the occult AVM in seven and partial removal of the lesion in one.

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