Vascular malformations of the brain stem

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Vascular malformations of the brain stem are unusual lesions that may pose a diagnostic and therapeutic challenge. Seven patients with vascular malformations involving the brain stem were evaluated; six were treated surgically, with complete obliteration of the lesion in five patients. In five patients symptoms developed only after a hemorrhage had occurred, and three of these suffered a rebleed before appropriate treatment was given. Angiography failed to demonstrate lesions in three cases, which did not appear to protect from repeat hemorrhage since two of the three rebled. There were no operative deaths, and no patients were made permanently worse after surgery. Useful recovery occurred commonly after appropriate treatment and appeared to be possible even in patients who had suffered a catastrophic neurological deficit at the time of presentation. These data indicate that surgical removal of the lesion may be warranted in some patients with symptomatic brain-stem vascular malformation.

KEY WORDS
arteriovenous malformation • subarachnoid hemorrhage • brain stem • brain-stem hematoma

VASCULAR malformations of the brain stem are unusual lesions and often present with catastrophic neurological dysfunction. Because of their treacherous location, surgical removal of these anomalies has been considered hazardous or impossible. Reports of resection of these lesions are rare and few authors recommend surgical treatment of brain-stem malformations.

Seven patients with vascular malformations involving the brain stem were referred to us for evaluation and treatment. The angioarchitecture of the lesion and the clinical presentation led to surgical treatment in six cases with acceptable management results. Surgical obliteration appears to be possible for some of these lesions and can be performed at an acceptable risk.

Summary of Cases

Clinical Presentation and Treatment

A summary of the clinical aspects of these seven patients is presented in Table 1. The age at onset ranged from 16 to 43 years (mean 32 years). Five patients were male and two were female.

Intracranial hemorrhage was the most common cause of presentation and occurred in five patients. Three patients, two with malformations limited exclusively to the brain-stem surface and one with a lesion involving the surface and the intra-axial region, presented abruptly with subarachnoid hemorrhage (SAH). Two other patients with strictly intra-axial malformations came to medical attention because of intrinsic brain-stem hematomas; in both instances, symptoms developed insidiously over several weeks. Because of the temporal profile of onset in these two patients, one was thought to have a brain-stem neoplasm and the other a demyelinating disease before the correct diagnosis was made at surgery. The remaining two patients presented with progressive brain-stem dysfunction not related to hemorrhage. This was thought to be due to "steal" in one case and to venous hypertension or local mass affect in the other.

All but one patient exhibited a profound neurological deficit that correctly localized the lesions to the brain stem. The one exception experienced an SAH not associated with localizing findings. Two patients (Cases 2 and 5) suffered respiratory arrest within 24 hours of hospital admission. Both of these patients made useful recoveries after appropriate therapy.

Angiography failed to demonstrate the malformation in three patients (in two of these, angiography was performed more than once). In all three patients the correct diagnosis was made at the time of surgery (two had strictly intra-axial malformations and one had a malformation limited exclusively to the brain-stem surface).

Preoperative computerized tomography (CT) and magnetic resonance (MR) imaging were performed on all patients and correctly demonstrated the location and extent of intra-axial involvement in five of the six
patients who underwent surgical exploration. In one patient (Case 1) the CT and MR studies were both interpreted as showing an intra-axial lesion of the tectum; however, surgical exploration revealed that the malformation was limited to the brain-stem surface.

Of the five patients who presented with hemorrhage, three experienced a repeat hemorrhage before appropriate treatment was given. Two of the three patients who suffered recurrent hemorrhages had normal angiograms.

Results of Treatment

Six of the seven patients underwent surgery. In five patients complete obliteration of the lesion was achieved. This was confirmed by postoperative angiography in patients in whom the malformation was visible on the preoperative angiogram. One patient (Case 6), with an intra-axial pontine malformation, required re-operation because the malformation was not identified or obliterated at the time of the first surgery. Another patient (Case 7) had a residual symptomatic malformation after endovascular embolization and was treated with surgical excision. All six patients who underwent surgery recovered; however, two had increased neurological deficits immediately following surgery. At their postoperative follow-up examination, five patients showed improvement over their preoperative status. The surgical patient who was worse (Case 3) deteriorated after gamma-beam irradiation, and her poor neurological function appears to be related to radiation rather than to her initial surgery. Three of the six patients who were treated surgically have returned to their premorbid occupations. Two of the three patients who were unemployed after surgery were not employed before their illness. All but one of these patients (Case 3) are sufficiently intact to be employable.

One patient (Case 4) with a mixed surface and intra-axial malformation was referred for stereotaxic Bragg peak proton beam irradiation. However, he was not treated because this did not appear to be financially feasible. He remains employed but continues to experience progressive neurological deterioration.

Representative Cases

Case 2

This 56-year-old right-handed man was well until 1 month before admission when he noted the onset of

TABLE 1

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age, Sex</th>
<th>Lesion Location</th>
<th>Type of Hemorrhage</th>
<th>Presentation</th>
<th>Angiographic Findings</th>
<th>Treatment</th>
<th>Follow-Up Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>36, F</td>
<td>quadrigeminal plate</td>
<td>SAH × 2</td>
<td>abrupt headache, Parinaud’s syndrome</td>
<td>normal × 2</td>
<td>supracerebellar, subtentorial resection</td>
<td>improved Parinaud’s syndrome; employed (field biologist)</td>
</tr>
<tr>
<td>2</td>
<td>56, M</td>
<td>lateral medulla</td>
<td>SAH × 1</td>
<td>abrupt headache, respiratory arrest</td>
<td>small malformation fed by PICA</td>
<td>retromastoid resection</td>
<td>neurologically normal; working (farmer)</td>
</tr>
<tr>
<td>3</td>
<td>21, F</td>
<td>4th ventricle &amp; brachium pontis</td>
<td>SAH × 2</td>
<td>abrupt headache, rt VI, VII spastic ataxic quadriparesis</td>
<td>malformation fed by penetrating arteries</td>
<td>suboccipital transvermian surgical exploration without resection, gamma-beam radiation</td>
<td>progressive brain-stem dysfunction beginning 6 mos after irradiation; not employed</td>
</tr>
<tr>
<td>4</td>
<td>35, M</td>
<td>midbrain</td>
<td>none</td>
<td>progressive tremor, diplopia, quadriparesis, ataxia</td>
<td>large AVM fed by basilar perforators</td>
<td>none</td>
<td>progressive deficit; employed (mechanic)</td>
</tr>
<tr>
<td>5</td>
<td>22, M</td>
<td>medulla</td>
<td>medullary hematoma × 1</td>
<td>subacute, ascending quadriparesis, ataxia, bilat V, VII, IX, X, XII; respiratory arrest</td>
<td>normal</td>
<td>suboccipital evacuation of hematoma &amp; resection of malformation</td>
<td>improved; mildly spastic ataxic gait &amp; dysarthria; not employed</td>
</tr>
<tr>
<td>6</td>
<td>42, M</td>
<td>pons</td>
<td>pontine hematoma × 2</td>
<td>subacute, progressive quadriparesis &amp; dysarthria</td>
<td>normal × 2</td>
<td>1: temporal, trans-tentorial evacuation of hematoma 2: repeat evacuation of hematoma &amp; resection of malformation</td>
<td>improved; mild dysarthria &amp; rt hemiparesis; employed (executive)</td>
</tr>
<tr>
<td>7</td>
<td>43, M</td>
<td>medulla</td>
<td>none</td>
<td>subacute headache, nausea, vomiting, hemiparesis, ataxia</td>
<td>dural fistulae w/ venous varix at cranio-cervical junction</td>
<td>1: embolization w/ PVA 2: suboccipital excision of recurrent malformation</td>
<td>improved; mild rt hemiparesis; not employed</td>
</tr>
</tbody>
</table>

* SAH = subarachnoid hemorrhage; PICA = posterior inferior cerebellar artery; AVM = arteriovenous malformation; PVA = polyvinyl alcohol. Roman numerals indicate cranial nerves affected.
† Age (years) at treatment.
‡ In Cases 1 and 2 the lesion was limited exclusively to the surface of the brain stem; in Cases 3 and 4 the lesion was intra-axial with involvement of the brain-stem surface; in Cases 5 and 6 the lesion was exclusively intra-axial; in Case 7 the lesion was predominantly a dural fistula.
Vascular malformations of the brain stem

intermittent double vision. On the day of admission he experienced the abrupt onset of headache.

On examination, he preferred to sleep but was easily aroused. His neurological examination was otherwise normal. A CT scan showed blood in the fourth ventricle and in the subarachnoid space around the brain stem. Shortly after admission, he became obtunded and suffered a respiratory arrest. He was intubated and a ventriculostomy was placed for cerebrospinal fluid drainage resulting in rapid improvement. Eventually, the ventriculostomy was converted to a ventriculoperitoneal shunt. Angiography showed a small vascular lesion filling from the left posterior inferior cerebellar artery (PICA).

At surgery, the left lateral medulla was approached through a left retromastoid craniectomy. The lesion itself was a "transistor"-shaped vascular malformation on the surface of the medulla. It was fed from pial branches rather than from the subarachnoid portion of the PICA that coursed past the lesion. The malformation was cauterized and removed. The patient tolerated surgery without neurological incident and made an uneventful recovery. At follow-up examination, he was neurologically normal and had returned to his previous occupation as a farmer.

Case 4

This 35-year-old left-handed mechanic presented with a 5-year history of progressive intention tremor involving the right arm and leg, diplopia, unsteady gait, right arm and leg weakness, and intermittent headache. He had no known episodes of intracranial hemorrhage.

On examination, he had a left medial longitudinal fasciculus deficit, marked intention tremor in both the right arm and right leg, and a mild right hemiparesis. He was able to walk with assistance but, in addition to his tremor, he had a spastic ataxic gait. Both plantar responses were extensor and he had generalized hyperreflexia. A CT scan (Fig. 1) and angiograms (Fig. 2) showed a large mixed surface and intra-axial malformation involving the midbrain. His progressive symptoms were believed to be due to arterial "steal." He was not judged to be a suitable candidate for surgery or endovascular embolization and was referred for Bragg peak proton beam irradiation; however, this treatment was not possible because of financial considerations. He continued to show progressive deterioration.

Fig. 1. Case 4. Contrast-enhanced computerized tomography scan showing an arteriovenous malformation involving the midbrain.

Fig. 2. Case 4. Left vertebral anteroposterior (left) and lateral (center) subtraction angiograms and left carotid subtraction angiogram (right) showing an arteriovenous malformation involving the upper brain stem.

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Case 6
This 42-year-old right-handed man had previously been healthy until he insidiously developed a progressive gait disturbance, tingling in the hands and feet, tremulousness, slurred speech, and right-arm weakness 6 weeks before admission. On examination he had mild dysarthria and right-sided hemiparesis involving the face, arm, and leg. Deep-tendon reflexes were hyperactive throughout and the right plantar response was extensor. His gait was ataxic. A CT scan showed a non-enhancing hypodense lesion in the pons, and MR imaging revealed a cystic lesion in the pons with a “fluid level” (Fig. 3). Angiography was normal.

A left subtemporal transtentorial approach was used to expose the pons, which appeared to be enlarged. The pons was opened revealing a cystic cavity filled with hemorrhagic fluid which was drained. No other abnormalities were found and several biopsy samples of the cyst wall were taken. Histological examination of these samples showed no evidence of abnormality. Immediately following surgery the patient’s right-sided weakness was somewhat worse, but this gradually improved. Progressive gait difficulties, dysarthria, tremulousness, tingling in the hands and feet, and right-sided weakness recurred several weeks after surgery. On neurological examination 8 weeks after surgery, his condition was somewhat worse than at his preoperative examination. Repeat CT and MR studies showed a recurrent hematoma in the pons. Angiography was again normal.

Reexploration of the pons was performed through the same exposure. The previous incision in the pons was enlarged and the hematoma evacuated. A cluster of tangled blood vessels was found in the wall of the hematoma cavity which was coagulated and removed. The patient’s condition was no worse following the second operation and thereafter it gradually improved. At his last follow-up examination, his only residual deficit was a mild right-sided hemiparesis and dysarthria. He has returned to his previous occupation as a business executive.

Case 7
This 43-year-old right-handed man had suffered transient episodes of severe right-sided headaches, nausea and vomiting, blurred vision, and transient right-arm weakness for several years before admission. Five days before admission he again developed severe right-sided headache, nausea and vomiting, and later the gradual onset of right-sided arm and leg weakness. Over the next several days his condition progressively worsened instead of improving as previously. He was noted to be lethargic at times. There was no history of trauma and he was otherwise well.

On examination, the patient was lethargic, but could easily be aroused. He had a moderate amount of weakness in the right arm and leg, worse distally than proximally, and the arm was weaker than the leg. He had generalized hyperreflexia, and both plantar responses were extensor. Sensation to pinprick and touch and proprioception were severely affected on the right side of the body. Testing of the cranial nerves and visual fields was normal. A CT scan with and without contrast enhancement did not show any evidence of hemorrhage, but revealed a large enhancing lesion at the craniocervical junction (Fig. 4). There was no hydrocephalus. A lumbar puncture was traumatic but not xanthochromic. Angiography revealed dural fistulae filling exclusively from the left external carotid artery with large venous varices surrounding the medulla (Fig. 5).

Endovascular embolic obliteration of the dural fistulae using polyvinyl alcohol was performed shortly after admission. Although he initially improved, hiccups and worsening right-sided weakness appeared 3 or 4 weeks after embolization. Repeat angiography showed that the dural fistulae were still patent, now filling not from the left external carotid circulation but via extradural branches of the left vertebral artery. Direct intra-
Vascular malformations of the brain stem

Dural arterial shunting into the venous varices was now apparent from the left anterior inferior cerebellar artery. Surgical exposure of the lesion was achieved through a midline suboccipital approach. The dural fistulae were identified and cauterized. Numerous small arterial channels from arterial pial vessels and larger vessels traveling in the subarachnoid space and fourth ventricle supplied arterial blood to the venous varices. These were cauterized and sectioned and the venous varices were removed. On the right side of the medulla, a varix was deeply embedded into the substance of the brain. Efforts to dissect this out of the brain stem were judged to be injudicious, and this varix was thrombosed with the neodymium:yttrium-aluminum-garnet (Nd:YAG) laser and an aneurysm clip was applied across its venous outflow.

Postoperatively, the patient developed transient bilateral vocal cord paralysis which required tracheostomy for a short period of time. His hemiparesis has gradually improved and at last follow-up examination was substantially better than at the time of admission. He is fully ambulatory and has no cranial nerve deficits at this time. Postoperative angiography showed no residual arteriovenous malformation.

Discussion

Vascular malformations of the brain stem are unusual lesions. The Cooperative Study found that 2% of all intracranial vascular malformations involve the brain stem. This series confirms the observation of others that brain-stem malformations most commonly come to medical attention only after intracranial hemorrhage has occurred. Young adults appear to be affected most often. Subarachnoid hemorrhage occurs more frequently than intra-axial hematoma; however, both are commonly accompanied by severe neurological dysfunction which usually localizes the lesion to the brain stem. Less frequently, brain-stem malformations produce progressive neurological dysfunction not related to identifiable intracranial hemorrhage. Although the risk of hemorrhage from asymptomatic lesions is unknown, these findings and those reported by others suggest that the risk of recurrent hemorrhage from symptomatic lesions in the absence of definitive therapy is substantial.

The presentation and treatment of patients in this series were related to the lesion angioarchitecture. Two patients (Cases 1 and 2) had malformations limited exclusively to the brain-stem surface. Both lesions received arterial blood from branches of the vertebrobasilar system at the pial surface or in the subarachnoid space and, as a consequence, the bulk of these lesions was limited to the pial surface of the brain stem or the subarachnoid space. Both manifested abruptly with SAH. Surgical excision was technically feasible and well tolerated. In two other patients (Cases 3 and 4), the malformations received arterial supply both from intra-axial penetration arteries and from surface arteries. Abrupt presentation with SAH occurred in one (Case 3). In two patients (Cases 5 and 6), the malformations were entirely intra-axial and were fed exclusively by penetrating branches of the vertebrobasilar system. Both of these lesions caused intra-axial hematoma leading to insidious, progressive brain-stem dysfunction. This temporal profile is not commonly associated with intracranial hemorrhage and may lead even the most astute clinicians away from the correct diagnosis.

Finally, in one patient (Case 7), arteriovenous shunting from the external carotid and extracranial vertebral arteries occurred in the dura and was associated with large intradural venous varices. Sources of arterial shunting from the intradural vertebrobasilar system became apparent after partial treatment. The angioarchitecture and presentation of this lesion were similar to those of spinal dural arteriovenous malformations.

Although angiography remains the cornerstone of diagnosis in patients with suspected intracranial vascular malformations, this series is unique in the high incidence of negative angiography. Failure to demonstrate the vascular malformation on preoperative angiography may occur because of small lesion size or because the lesions are not filling at the time of angiography. Regardless of the cause, in this series rebleeding occurred at the same high incidence both in patients whose vascular malformations could not be demonstrated on angiography and in those with angiographically demonstrable malformations. Decisions regarding therapeutic intervention, therefore, cannot be based exclusively on whether a vascular malformation is seen on angiography. Both CT and MR studies complement angiography and better define the anatomy of these lesions. In particular, the extent of intra-axial malformations can often be predicted based on the MR or CT images. Despite these high-resolution studies, the operative findings may differ from those predicted.

The results of this series show that, despite a catastrophic neurological presentation, useful recovery is common after appropriate treatment. In this series, even patients who presented with respiratory paralysis eventually recovered. Overall, six of the seven patients were treated surgically and complete obliteration of the vascular malformation was achieved in five patients.
There were no deaths, and no significant morbidity was related to surgical intervention. Patients with small intra-axial malformations tolerated surgical resection without additional permanent neurological deficits. These results, which are somewhat better than those reported by others, most likely reflect patient differences rather than fundamental variations in surgical technique.\textsuperscript{1,5,6,23} Surgery appears to be warranted in some patients with brain-stem vascular malformations. Although some malformations may lend themselves to surgical resection, injudicious efforts to resect lesions with extensive intra-axial components may result in disaster.\textsuperscript{6} For these malformations, some success has been reported with high-energy irradiation; however, as our Case 3 illustrates, this mode of treatment is not without risk.\textsuperscript{25} Radiation therapy does appear to lower the long-term danger of repeat hemorrhage, but this effect is not immediate and may leave the patient unprotected for several years. Little is known about the long-term consequences of this type of treatment. Endovascular embolization is usually not possible because of the anatomical and flow characteristics of these lesions. Surgical resection should be considered for symptomatic brain-stem malformations before other modes of treatment are explored.

References


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