Diagnosis and treatment of vascular brain-stem malformations

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Vascular malformations are a common cause of spontaneous brain-stem hemorrhage in young normotensive individuals. These lesions are no longer cryptic. Magnetic resonance (MR) imaging has renewed interest in the treatment of this disorder because of the precise accuracy in diagnosis and localization of these lesions that it affords. The MR image demonstrates characteristic findings of multiple hemorrhages of varying ages surrounded by a hypointense peripheral zone of hemosiderin. Five cases of vascular brain-stem malformation diagnosed with MR imaging are described. The vascular malformations could be demonstrated as "flow void" areas in three cases. Three patients were treated surgically and vascular malformations were confirmed; all three patients improved postoperatively. Two patients were treated nonsurgically; one of these recovered from a second hemorrhage and the other experienced neurological deterioration after a single hemorrhage. High-energy radiotherapy was not effective for the one vascular malformation treated by this method. This experience suggests that surgical exploration should be considered for vascular brain-stem malformations when the diagnosis is confirmed by MR criteria and the clinical course and lesion are both progressive in character.

KEY WORDS: vascular malformation, pontine hematoma, brain stem, magnetic resonance imaging

The recent development of magnetic resonance (MR) imaging has renewed interest in vascular malformations of the brain stem.1,3,14,21,36 Pathological studies indicate that the pons is one of the most common sites of these malformations.26,27,48 Their association with primary pontine hemorrhage in young, normotensive individuals makes premorbid recognition vitally important. Vascular malformations have been confirmed histologically in autopsy cases of pontine hematoma23,38,43 and in several cases of surgically evacuated pontine hematoma.7,12,33,40,47,51 Preoperative diagnosis has been difficult due to poor resolution of computerized tomography (CT) scans in the retroclival region.3,5,29 Total surgical removal has not been documented prior to this report.

The clinical diagnosis is frequently obscure. These malformations may present a course that mimics neoplastic disease,10,23,28,32,41 demyelinating disease,1,2,7,20,23,42 and acute and subacute cerebral infarction.10,19,30 The natural history of these lesions is poorly documented. Most reported cases present with signs and symptoms of progressive brain-stem deterioration characterized by recurring hemorrhage. The end-point of the follow-up period for patients in these series is major disability,1,24,26,43 surgical treatment,4,5,11,15,18,19,23,28,30,34,36,40,41,44,50,51 or death.

We present five cases of pontine hematoma due to vascular malformations which were not visualized by angiography but were diagnosed by MR imaging. The patients ranged in age from 22 to 36 years. There were three females and two males. The multiplanar imaging capability of the MR technology provides accurate localization of brain-stem lesions and documents a surgical corridor to the lesion which minimizes operative trauma.

Case Reports

Case 1

This 25-year-old left-handed man had been well until 1 month prior to admission, when he noted the sudden onset of paresthesias and incoordination of the left hand. These symptoms progressed over several days with development of diplopia on right gaze and weakness of the left upper extremity.
FIG. 1. Case 1. Magnetic resonance images, sagittal T1-weighted (spin-echo, TR 500/TE 20 msec) image (left) and axial T2-weighted (spin-echo, TR 2000/TE 80 msec) image (right), showing signal changes of acute hemorrhage (deoxyhemoglobin) and subacute hemorrhage (intracellular and free methemoglobin) within the pons. Hypointense nodule (arrow, left) anterior to the pons, probably represents hemosiderin from a remote hemorrhage. Curvilinear hypointensity (arrow, right) is believed to represent a "flow void" area within a vessel.

Examination. Pertinent findings on neurological examination consisted of right abducens nerve palsy, hypesthesia and hypalgesia over the left side of the face, bilaterally diminished gag reflex, mild weakness of the left upper extremity, diminished fine motor skills, and a left Babinski reflex. Contrast-enhanced CT showed multiple abnormal vessels in the lateral walls of the fourth ventricle. Angiography demonstrated abnormal vessels in the venous phase representing a venous angioma. An MR study showed a 1.5-cm pontomedullary mass with signal changes characteristic of subacute and acute hemorrhage. Adjacent to the mass were multiple serpiginous signal-void areas, indicating flowing blood in aberrant vessels (Fig. 1).

Operation. Surgical exploration was performed with the patient in the lateral-oblique position. A midline suboccipital craniotomy was created with section of the inferior cerebellar vermis. Two large veins were identified to the right of midline on the floor of the fourth ventricle at the level of the facial colliculi. The vein entered the floor of the fourth ventricle lateral to the median raphe. These large veins were coagulated with bipolar cautery and the brain stem was entered between them. A hematoma was evacuated and several smaller veins were coagulated. No attempt was made to totally excise the hematoma en mass, but all microscopic vessels were obliterated with bipolar coagulation.

Postoperative Course. The postoperative neurological examination documented the addition of right facial paralysis and gait ataxia to the preoperative deficit. Neurological function improved to allow the patient to return to his employment as a computer technician. A hypoglossal-facial nerve anastomosis was performed to reanimate the face. The patient has been followed for 24 months. There has been no evidence of recurrent hemorrhage and a recent MR image demonstrated no evidence of residual malformation.

Case 2

This 22-year-old right-handed woman developed paroxysmal right frontal headache on August 23, 1987, followed by paresthesias of the right side of the face and body and horizontal diplopia on right gaze.

Examination. Pertinent findings on neurological examination consisted of horizontal nystagmus on right gaze, hypesthesia and hypalgesia over the right side of the face and body, dystnesia in the right upper extremity, and a wide-based unsteady gait. A CT scan demonstrated a 2.0-cm pontine hematoma which was associated with displacement of the floor of the fourth ventricle. Angiography was normal. Magnetic resonance imaging demonstrated a mass in the pons with signal changes characteristic of mixed acute, subacute, and chronic hemorrhage. A halo of low signal consistent with the presence of hemosiderin surrounded the hematoma (Fig. 2 left and center).

The patient's symptoms resolved almost completely over several days. She continued to do well until 2 months later, when she suffered another episode of frontal headache, followed by increasing sensory deficit.
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of the right side of the face and body, diplopia, and incoordination of the right hand. Her gait became unsteady, and she required support. Neurological examination documented horizontal nystagmus, partial right abducens nerve palsy, mild right pronator muscle drift, hypesthesia and hypalgesia over the right side of the face and body, dysmetria and incoordination of the right upper and lower extremities, and a wide-based unsteady gait with falling to the right. Mild hyperreflexia was noted in all extremities. An MR study showed an increase in the size of the pontine hematoma with new signal characteristics consistent with acute hemorrhage.

Operation. Surgery was performed with the patient in a lateral-oblique position. A midline suboccipital craniotomy was carried out with section of the inferior cerebellar vermis (Fig. 3). The lateral aspect of the floor of the fourth ventricle was bulging posteriorly beneath an area of bluish discoloration. The hematoma cavity was opened and dark-brown fluid was evacuated. A mass consisting of hematoma and small blood vessels was totally excised with bipolar cautery and laser.

Postoperative Course. The postoperative neurological examination showed partial internuclear ophthalmoplegia and increased gait ataxia. Histological evaluation of the excised specimen demonstrated a venous angioma (Fig. 4). Postoperative MR imaging confirmed that the lesion was totally removed (Fig. 2 right). The patient recovered neurological function and has subsequently returned to her previous employment as a primary-school teacher.

Fig. 2. Case 2. Left: Preoperative sagittal T1-weighted (spin-echo, TR 500/TE 20 msec) magnetic resonance (MR) image demonstrating a signal characteristic of subacute hemorrhage (arrow) within the pons. Center: Preoperative axial T2-weighted (spin-echo, TR 2000/TE 80 msec) MR image revealing an increased signal characteristic of subacute hemorrhage (free methemoglobin). A thin rim (arrow) of hypointensity represents hemosiderin-laden macrophages. Right: Postoperative axial T1-weighted (spin-echo, TR 500/TE 20 msec) MR image demonstrating complete resection of malformation and hematoma.

Fig. 3. The patient is operated on in a lateral oblique position as demonstrated in the drawing (lower). A midline incision and suboccipital craniotomy are performed to provide access to the cerebellar vermis. Under the operating microscope the vermis is divided and self-retaining retractor blades are placed to provide access to the floor of the fourth ventricle. The malformation and hematoma are resected with impedance-monitored bipolar coagulation and the neodymium: yttrium-aluminum-garnet (Nd:YAG) laser.
Case 3

This 36-year-old man developed the sudden onset of severe headache.

Examination. Neurological examination revealed right facial palsy, right abducens nerve palsy, anesthesia of the left side of the body, decreased coordination of the left extremities, left hyperreflexia, and left Babinski reflex. A CT scan demonstrated a pontine mass which was surgically explored through a suboccipital craniectomy at another institution. Subsequently, MR imaging revealed a 2.0-cm pontine mass with a signal characteristic of acute hemorrhage surrounded by a thin rim of low signal due to hemosiderin (Fig. 5 left). Angiography was normal.

The patient received high-energy proton-beam radiotherapy for a presumptive diagnosis of cavernous angioma. Subsequent to this treatment, he was observed closely for 2 years. He continued to demonstrate sequences of deterioration of cranial nerve, cerebellar, and left pyramidal function. An MR image demonstrated an expanding mass lesion consisting of hematomas of varying ages.

Operation. Surgery was performed with the patient in the lateral-oblique position. A midline suboccipital craniotomy was made with section of the inferior cerebellar vermis. The right lateral aspect of the floor of the fourth ventricle was replaced by a grape-like mass in the region of the facial colliculus (Fig. 6). The cavity was opened and the malformation and hematoma were extirpated from the brain stem.

Postoperative Course. The patient’s neurological examination demonstrated a left partial sixth nerve palsy and left peripheral facial palsy in addition to the preoperative neurological findings. He continues to improve to a condition considerably better than his preoperative status. Histological examination of the surgical specimen demonstrated a venous angioma. Postoperative MR imaging demonstrated complete resection of the malformation and hematoma (Fig. 5 right).

Case 4

This 34-year-old right-handed woman had been well until 10 days prior to admission, when she developed bitemporal headache. The following morning she continued to have bitemporal and occipital headache, and developed a fluctuating horizontal diplopia on right gaze. Over several days she developed motion sickness, patchy paresthesias of the left upper and lower extremities, clumsiness of the left hand, dysarthria, and dysphagia.

Examination. Pertinent findings on neurological examination consisted of right abducens nerve palsy, right lower facial weakness, decreased right auditory acuity, left hemiparesis, and left Babinski reflex. Computerized tomography demonstrated a 2.0-cm pontine hematoma (Fig. 7 lower right). Angiography did not
Vascular brain-stem malformations demonstrate any abnormal vessels. An MR study showed a pontomedullary mass with the signal characteristics of mixed acute and subacute hemorrhage. The mass was surrounded by an increased signal characteristic of edema (Fig. 7 upper). No abnormal vessels were visible in the brain stem; however, incidental findings of multiple foci of hypointensity in the cerebral white matter were thought to represent small vascular malformations containing chronic hemorrhage (Fig. 7 lower left).

**Course.** The patient’s symptoms gradually improved over the following month. A moderate degree of incoordination of the left upper extremity and left abducens nerve palsy persisted. Three months later, she suffered a repeat bout of headache and worsening of her neurological condition. A CT scan showed recurrent pontine hemorrhage. She recovered and has returned to her position as an executive secretary. Surgical treatment is not planned unless progressive deterioration is demonstrated.

**Case 5**

This 36-year-old right-handed man developed the abrupt onset of dizziness, nausea and vomiting, and vertigo 3 days prior to admission. On the day before admission, he experienced right facial numbness and difficulty in swallowing.

**Examination and Course.** The neurological findings included nystagmus on right lateral gaze, decreased facial sensation and corneal reflex on the right, and an ataxic gait. A CT scan demonstrated a 1.0-cm pontine hematoma. An MR study showed a pontomedullary mass with the signal characteristics of acute hemorrhage (Fig. 8). The mass was surrounded by an increased signal characteristic of edema. A signal flow-void area consistent with vascular malformation was present within the hematoma. Angiography was normal. Over
several days, the patient’s neurological examination returned to normal. He has returned to his employment as a newspaper editor and is being observed for evidence of recurrent hemorrhage.

Discussion

The term “cryptic” vascular malformation was introduced in 1956 by Crawford and Russell to designate small vascular malformations which escaped angiographic detection but were identified histologically in cases of spontaneous cerebral hemorrhage. Cryptic malformations represent a heterogeneous group of vascular malformations with variable histological characteristics, and include cavernous angiomas, venous angiomas, telangiectasias, and totally thrombosed arteriovenous malformations. Computerized tomography has improved detection of brain-stem lesions but lacks the specificity to differentiate vascular malformations from other lesions such as tumors and granulomas. Magnetic resonance imaging has increased sensitivity and specificity in the diagnosis of these lesions and can differentiate between acute hemorrhage (intracellular deoxyhemoglobin), subacute hemorrhage (intracellular and free methemoglobin), and chronic hemorrhage (hemosiderin); evaluation by MR imaging of such vascular malformations demonstrates a relatively uniform and characteristic morphology, so it is difficult to call them “cryptic.” The typical findings are a central focus of abnormal signal indicating acute hemorrhage associated with intracellular deoxyhemoglobin (hypointense to isodense with brain on T1-weighted images, hypointense on T2-weighted images), subacute hemorrhage with intracellular and free methemoglobin (hyperintense on T1-weighted images, hyperintense on T2-weighted images), or chronic hemorrhage with hemosiderin (extremely hypointense on both T1- and T2-weighted images).

Multiple hemorrhages of varying ages are common in occult malformations and the aging quality of hemoglobin permits accurate differentiation by MR imaging. Occasionally a signal-void area caused by blood flowing in vessels can be detected inside or adjacent to the hemorrhage. In the patients reported here, central foci consisted of subacute hemorrhage (Cases 1 and 3) and multiple hemorrhages of different ages (Cases 2 and 4). Acute hemorrhage was seen in Case 5. All patients showed peripheral hypointense rims indicating the presence of (chronic) hemosiderin. Vascular malformations were demonstrated as “flow void” areas in Cases 2, 3, and 4.

The age of the hemorrhages shown on MR imaging correlated well with the timing of the clinical signs of hemorrhage in most patients. In two patients who underwent surgery, a venous angioma was proven histologically. In the two nonsurgical patients, histological confirmation has not been obtained. In contrast to the acute serious course of hypertensive pontine hemorrhage, pontine hematoma due to rupture of a vascular malformation presents a relatively milder onset and variable clinical course. The most common presentations are: progressive neurological deterioration over days or weeks mimicking pontine glioma, relapsing and remitting course lasting over a period of weeks, months, or even years suggesting demyelination, or subacute onset suggesting an infectious process or stroke of the brain stem.

There are numerous isolated reports of successful evacuation of a pontine hematoma in the literature. The most commonly utilized surgical procedure is a posterior approach through the inferior vermis, exposure of the lesion in the floor of the fourth ventricle, and drainage of the hematoma. Supracerebellar approach to the upper pons, subtonsillar approach to the lower pons, and subtemporal approach with pedunculotomy have also been reported. Stereotactic evacuation of a brain-stem hematoma is an accepted method of treatment; however, vascular malformations may not be adequately treated or diagnosed by this procedure. Total removal of the hematoma and malformation is the preferred form of treatment.

The outcome after surgical evacuation of pontine hematomas has been generally good. In most cases, neurological deficits were reported to be improved or stabilized. There were only a few cases in which patients suffered severe or progressive neurological deterioration. The natural history of pontine hematoma due to rupture of vascular malfor-
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In most cases, signs and symptoms showed progressive or fluctuating deterioration over weeks, months, or years until either surgery or death intervenes. Recurrent hemorrhage with neurological deterioration is documented clinically. Pathological examination of autopsy cases and MR imaging documents multiple hemorrhages of different ages in most cases. External irradiation has been proposed for treatment of cavernous angiomia of the brain, however, its efficacy has not been proven in other types of cryptic vascular malformations (L. Steiner: personal communication, 1989). We have observed a patient with brain-stem vascular malformation who developed slowly progressive neurological deterioration after radiation therapy. Sano and Ochiai compared surgical treatment with medical treatment, and concluded that pontine hematomas in nonhypertensive patients under 40 years of age should be explored in an effort to extirpate the suspected cryptic vascular malformation and prevent rebleeding.

The present experience documents the ability of MR imaging to diagnose heretofore "cryptic" vascular malformations of the brain stem. In view of this greater diagnostic accuracy, the precise anatomical localization, and the acceptable surgical morbidity compared with the morbidity associated with nonsurgical therapy, surgical extirpation for lesions associated with progressive or recurrent neurological deterioration is clearly recommended. The location of the lesion in surgically accessible sites may mandate surgical treatment in patients who have not suffered major disability. Careful evaluation of the natural history and the course of results following surgical and radiation therapy is needed to determine the ultimate choice of treatment for early detected lesions.

Addendum

Since submission of this manuscript, the authors have evaluated and treated six additional patients with vascular brain-stem malformations. Two patients remain neurologically intact after a single hemorrhage. Four more patients presented with progressive neurological deficits secondary to multiple hemorrhages. These were three women and one man, ranging in age from 23 to 43 years. These four patients underwent complete surgical extirpation of a vascular malformation and hematoma; histological evaluation demonstrated cavernous angiomia in all cases. Only one patient exhibited additional morbidity related to hydrocephalus after removal of a malformation from the mesencephalon. The other three patients have returned to gainful employment.

This additional experience further documents the necessity of surgical treatment for vascular brain-stem malformations.

References


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