Cryptic vascular malformations of the spinal cord: diagnosis by magnetic resonance imaging and outcome of surgery

STANLEY L. BARNWELL, M.D., PH.D., CHRISTOPHER F. DOWD, M.D., RICHARD L. DAVIS, M.D., MICHAEL S. B. EDWARDS, M.D., PHILIP H. GUTIN, M.D., AND CHARLES B. WILSON, M.D.

Departments of Neurological Surgery, Radiology, and Pathology, School of Medicine, University of California, San Francisco, California

The cases of seven patients with intramedullary, cryptic vascular malformations of the spinal cord are reported. In all patients, the clinical course was progressive; a Brown-Séquard syndrome was the most common presenting symptom complex. Magnetic resonance (MR) imaging was performed in all patients. The pattern seen most often was a focus of high signal (on both T1- and T2-weighted MR images) surrounded by a larger zone of low signal (best seen on T2-weighted images), and was remarkably similar for all patients. Six patients underwent surgical exploration; removal of the lesions halted the progression of symptoms in five patients, and one patient had worsened sensory function after surgery. Motor function did not decrease postoperatively in any patient. The one patient who refused surgery has continued to decline neurologically. Histopathological examination of surgical specimens showed a cavernous malformation in one patient, a venous malformation in one, varices in two, and organizing hematomas in two; these findings are markedly different from those in previously reported cases of cryptic vascular malformations.

KEY WORDS • vascular malformation, cryptic • spinal cord • magnetic resonance imaging

Cryptic vascular malformations are small lesions that are difficult to detect during routine angiography. The term “cryptic vascular malformation” does not denote a specific size or histology. These lesions are frequently found in the brain, but their occurrence in the spinal cord is extremely rare. It is possible to detect these lesions with magnetic resonance (MR) images of the spinal cord. The natural history, histopathology, and outcome of treatment for seven patients with MR-documented cryptic vascular malformations of the spinal cord are reported here.

Clinical Material and Methods

Patient Characteristics

This series included four males and three females, ranging in age from 9 to 60 years (Table 1). Five patients were between 30 and 40 years of age. The medical history of one patient (Case 3) was of interest because he had undergone resection of a spinal arteriovenous malformation (AVM) 29 years before being evaluated for a cryptic lesion of the spine.

Presentation

All patients presented with sensory dysfunction, including numbness or pain that was often acute in onset; three patients had a Brown-Séquard syndrome. All patients described their symptoms as being progressive, although progression was often stepwise. The progressive symptoms included worsening pain, numbness, weakness, or spasticity. One patient (Case 5) was in her second trimester of pregnancy when her symptoms worsened acutely. The duration of symptoms varied from 4 weeks (Cases 4 and 5) to over 10 years (Case 3).

Radiological Evaluation

Magnetic resonance imaging was performed in all patients, and typically showed an intramedullary mass lesion that extended over one to three vertebral segments. A central area of high signal intensity, consistent with the presence of a methemoglobin, was seen on both T1- and T2-weighted images. Surrounding zones of signal hypointensity, best seen on the T2-weighted images, were compatible with the presence of hemosi-
Clinical summary of seven cases of cryptic vascular malformation (CVM)

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex, Age (yrs)</th>
<th>Location of CVM</th>
<th>Deficits on Presentation</th>
<th>Duration of Symptoms</th>
<th>Angiographic Findings</th>
<th>Removal of Lesion</th>
<th>Pathology</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M, 37</td>
<td>T6–7</td>
<td>Brown-Séquard</td>
<td>3 yrs</td>
<td>not done</td>
<td>complete</td>
<td>cavernous malformation</td>
<td>stable</td>
</tr>
<tr>
<td>2</td>
<td>M, 40</td>
<td>C-2</td>
<td>sensation</td>
<td>6 mos</td>
<td>venous pooling</td>
<td>partial</td>
<td>hematoma</td>
<td>stable</td>
</tr>
<tr>
<td>3</td>
<td>M, 32</td>
<td>T5–6</td>
<td>sensation</td>
<td>10 yrs</td>
<td>negative</td>
<td>partial</td>
<td>hematoma</td>
<td>stable</td>
</tr>
<tr>
<td>4</td>
<td>F, 9</td>
<td>C-6</td>
<td>Brown-Séquard</td>
<td>4 wks</td>
<td>negative</td>
<td>complete</td>
<td>venous varix</td>
<td>improved</td>
</tr>
<tr>
<td>5</td>
<td>F, 30</td>
<td>T-6</td>
<td>sensation</td>
<td>4 wks</td>
<td>not done</td>
<td>complete</td>
<td>venous malformation</td>
<td>worse sensation</td>
</tr>
<tr>
<td>6</td>
<td>F, 38</td>
<td>T5–6</td>
<td>sensation, spastic</td>
<td>15 wks</td>
<td>negative</td>
<td>complete</td>
<td>venous varix</td>
<td>improved</td>
</tr>
<tr>
<td>7</td>
<td>M, 60</td>
<td>T-4</td>
<td>Brown-Séquard</td>
<td>6 yrs</td>
<td>not done</td>
<td>none</td>
<td>none</td>
<td>worse</td>
</tr>
</tbody>
</table>

derin. No abnormal vessels were identified. The spinal cord was expanded at the location of the lesion in a nonspecific manner. Cystic changes or a syrinx were occasionally found adjacent to the mass lesion.

Spinal angiography with selective catheterization of intercostal arteries was performed in four patients. An abnormal vascular blush was seen in only one patient (Case 2). No other vascular abnormalities were observed.

**Surgery**

Surgical exploration was performed in six patients; the seventh patient deferred surgical therapy. The gross surgical findings were similar in all cases. After exposure of the dura over the lesion through a laminectomy that encompassed a level above and below the mass, intraoperative ultrasonography was used to localize the lesion precisely. The mass was exposed by opening the dura in the midline under the operating microscope; the mass was most often a dark, purple hematoma situated subpially within the cord. In all cases, the subarachnoid space was clear and without signs of hemorrhage. In a few patients, arterialized veins were seen overlying the lesion, and a thrombosed vein was observed in one patient. The mass was opened by incising through the pia into the cord; the mass was filled with hematoma of various consistencies, including fresh hematoma and older, organized hematoma. After the hematoma had been removed gently with suction irrigation, small blood vessels, some of which were thrombosed, were seen along the wall of the hematoma cavity. Affected vessels were either coagulated or removed. The extent of removal of the hematoma and abnormal vessels depended on the adherence of the mass to the adjacent spinal cord parenchyma, and was thought to be complete in four of the six cases. Material for pathological examination was obtained from all six surgical patients.

**Results**

**Outcome of Surgery**

Immediately after surgery, all patients experienced worsened symptoms, including numbness and decreased proprioception. With the exception of one patient (Case 6), whose spastic bladder condition worsened, no patient suffered significantly altered motor or sensory function after the perioperative period. The exception (Case 6) exhibited recurrent weakness 3 weeks after surgery. In that patient, an MR image showed bloody fluid within the surgical site; after treatment with dexamethasone, function gradually improved and eventually surpassed the preoperative function.

**Follow-Up Results**

All patients were followed for between 2 months and 2 years. At a 2-month follow-up examination, the increased numbness and worsened spastic bladder in Case 5 had not improved. Five patients, including the patient with a postoperative hemorrhage, noted a decrease in pain and an improvement in sensation; their symptoms had not been exacerbated by the surgery. The patient who declined surgery (Case 7) has continued to deteriorate slowly, with an increase in pain and spasticity.

**Histopathological Findings**

Examination of the operative specimens from all six patients undergoing surgery showed organizing hematoma and hemosiderin. Histopathological evidence of a cavernous malformation was found for Case 1. There was evidence of multiple venous channels consistent with a venous malformation in Case 5 and with a venous varix in Cases 4 and 6. Gliosis was noted in the two samples of neural tissue obtained at surgery. There was no histopathological evidence of an AVM in any specimen examined.

**Illustrative Case Report**

This 37-year-old man (Case 1) presented with a 3-year history of low-back pain that also involved his legs and chest. He also complained of dysesthesias involving his trunk and legs. During the year before diagnosis was made, he developed spastic paraparesis.

**Examination.** His general examination was unremarkable. On neurological examination he was found to have a Brown-Séquard syndrome, with decreased pinprick sensation on the right and moderate leg weakness on the left. He had bilateral Babinski signs and
Cryptic vascular malformations of the spinal cord

hyperreflexia. An MR image showed a well-circumscribed 1-cm mass in the dorsal aspect of the spinal canal at T-6 (Fig. 1). Compared with normal spinal cord, the mass had a slightly increased MR signal on both short and long repetition time (TR) sequences. The short TR images also showed an ill-defined region of hyperintensity within the spinal cord cephalad to the mass, which is most consistent with a subacute hemorrhage. Preferential shortening surrounding the mass on the long TR T2-weighted images indicated the presence of hemosiderin. Spinal angiography was not performed because MR imaging did not show the presence of abnormal vessels.

Operation. A thoracic laminectomy was performed from T-6 through T-9. The epidural space and dura were normal in appearance and there was no abnormal vasculature. Ultrasound studies were used to localize the highly echogenic lesion. The dura was opened in the midline at the level of T-6. A bulging, largely thrombosed vascular malformation, covered only by tightly stretched glistening white strands of pia mater, was seen on the left side of the dorsal surface of the spinal cord (Fig. 2). Red vessels crossed the surface of the cord from right to left at that level, and a collection of two coiled red vessels passed rostrally almost at the midline. Upon coagulation with microbipolar forceps, it was evident that these were arterialized veins. The dentate ligament was detached and, with the aid of a fine stay suture, the cord was rotated in order to observe the ventral lateral surface of the malformation. The cord was yellow-brown in color beneath the malformation. At least six very small arteries entered the malformation from its ventral aspect. The malforma-

Postoperative Course. Postoperatively, the patient had increased weakness of his left leg and decreased sensation in his right leg. Over several days, these symptoms began to resolve. At the time of discharge 10 days later, he was able to ambulate with the aid of a walker. His major complaint was a decreased proprioception.

Pathological Examination. Light microscopic examination of the tissue showed numerous venous channels indicative of a cavernous hemangioma (Fig. 3).

Discussion

Cryptic vascular malformations of the brain have been reported frequently and their clinical course, pathology, and treatment have been well described. The presence of similar lesions in the spinal cord has been reported only occasionally, and little is known about the disease process.

In a recent review of cryptic vascular malformations in the brain, Lobato, et al., reported that AVM’s and cavernous malformations are the most commonly identified vascular anomaly, accounting for 75% of these lesions. Venous malformations were found in only 10% of cases. The existence of mixed forms of vascular malformations indicated that pathological classification may not be of practical importance. The clinical presentation was similar among the various forms of vascular malformations.

In our patients, the natural history of cryptic vascular malformation of the spinal cord was characterized by progressive neurological dysfunction that occurs in either a continuous or a stepwise fashion. Sensory tracts were always affected, and a Brown-Séquard syndrome was often found. The majority of the signs and symptoms are related to hemorrhage, with acute exacerbations of pain or neurological deficits. Most patients, however, noted a gradual progression of either pain,
numbness, or weakness, the course of which was not clearly related to major bleeding. Because most patients improved when given dexamethasone, some component of edema may have been present, and T2-weighted MR images showed some edema. Syrinx or cyst formation associated with the lesion was occasionally seen, and this may have contributed to neurological deterioration.

The availability of MR imaging has made possible the diagnosis of cryptic vascular malformation of the
Cryptic vascular malformations of the spinal cord

In our patients, MR images showed a focal intramedullary abnormality, usually 1 to 3 cm in size, that had signal characteristics consistent with hemorrhage of variable age. In some patients, there were associated cystic areas adjacent to the focal lesion that were usually filled with blood. Serpentine vessels often found in spinal AVM's were not seen. Computerized tomography and myelographic findings were nonspecific.

Angiography was not necessary when the MR image was diagnostic. Some abnormal venous pooling was observed on the angiograms of one patient, but in general this test was not helpful. Spinal angiography should not be used routinely to evaluate spinal hematomyelia when a cryptic vascular malformation has been diagnosed.

Venous malformations that caused hemorrhage were well defined in one patient (Fig. 4). This lesion consisted of numerous veins that appeared to be normal; they were visualized best with a trichrome stain. Venous varices were identified in two other patients. In one of the two patients in whom only an organizing hematoma was found, an operation for a purported spinal AVM had been performed 29 years before the diagnosis of the current condition was made. The pathological specimen from the previous operation was not available, so a relationship between his initial lesion and his present problem cannot be ascertained.

Recently, Cosgrove, et al., reported a series of five cases of cavernous malformation of the spinal cord presenting as a cryptic vascular malformation. These cases were very similar to ours, both in the clinical course and the appearance of the MR images. The histological specimens upon which the diagnosis of cavernous angioma was made were not shown. With the possible exception of Case 1, the lesions described in the present report were not cavernous angiomas.

The surgical treatment of these lesions effectively prevented further deterioration. In four of six patients, it was possible to remove the lesion almost completely. In two patients, subtotal resection was performed because of the dense adherence of the lesion to the adjacent cord. Surgery may worsen the sensory changes, but with time these changes have resolved in this series. Five of six patients who underwent surgery have noted improvement from their preoperative condition, and no patient had decreased motor function after the operation. In one patient, hemorrhage occurred into the operative site, but all effects related to this postoperative complication resolved. Removal of the cryptic vascular malformation appeared to protect the patient from further deterioration.

References


Manuscript received March 23, 1989. Accepted in final form August 28, 1989.

Address reprint requests to: Stanley L. Barnwell, M.D., Ph.D., c/o The Editorial Office, 1360 Ninth Avenue, Suite 210, San Francisco, California 94122.